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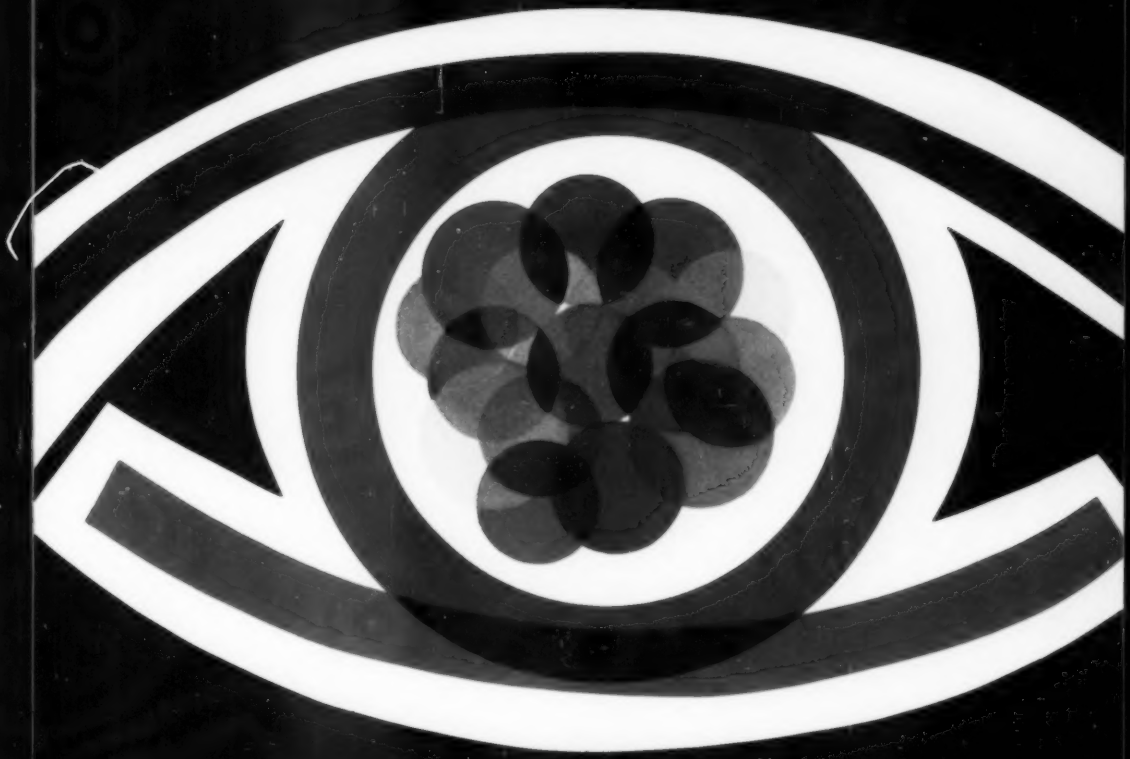

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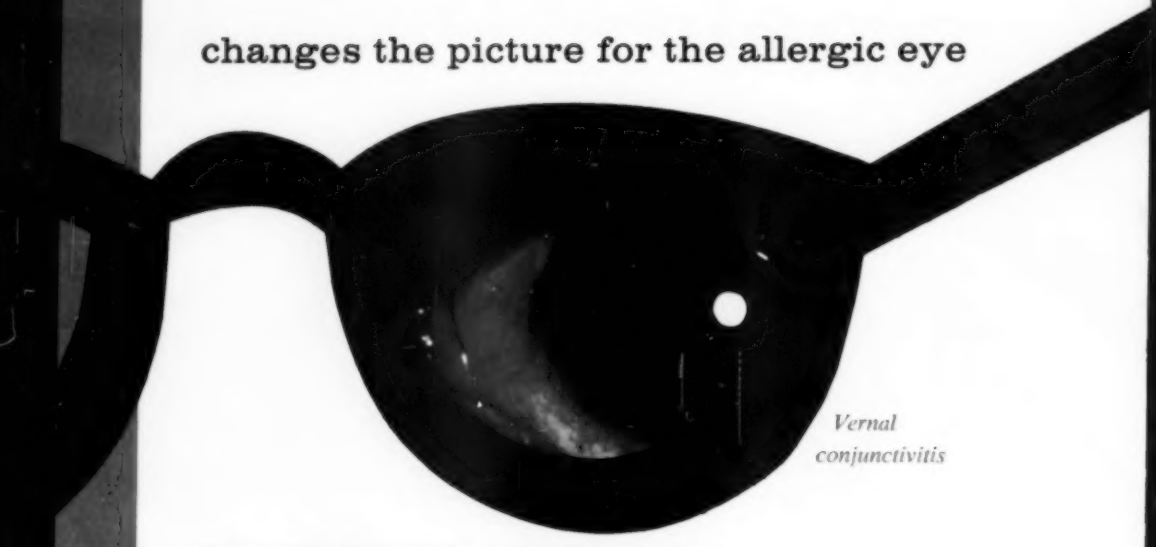

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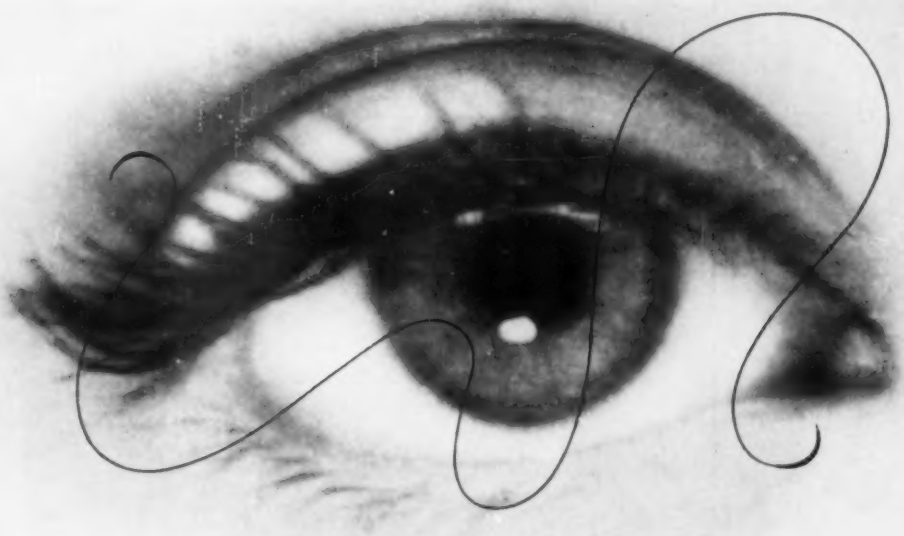

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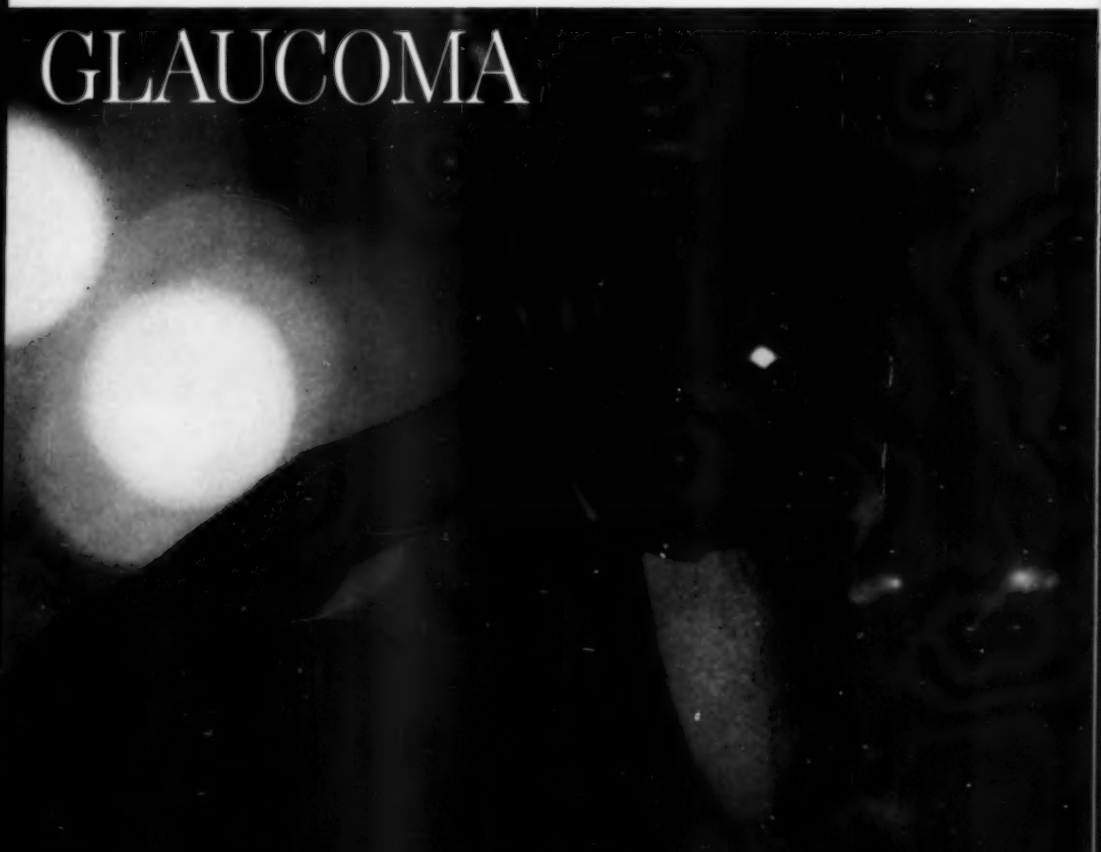

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22	23	24	25	26	27	28
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REFERENCES: (1) Perkins, E. S.: *Practitioner* 178:875, 1957. (2) Queries and Minor Notes, *J.A.M.A.* 161:1032, 1956. (3) Smith, C. H.: *Eye, Ear, Nose & Throat Month.* 34:580, 1955. (4) Blakiston's New Gould Medical Dictionary, ed. 2, New York, McGraw-Hill Book Company, Inc., 1956, p. 945. (5) Cutler, H. B., & Braley, A. E.: *J. Iowa M. Soc.* 44:427, 1954.

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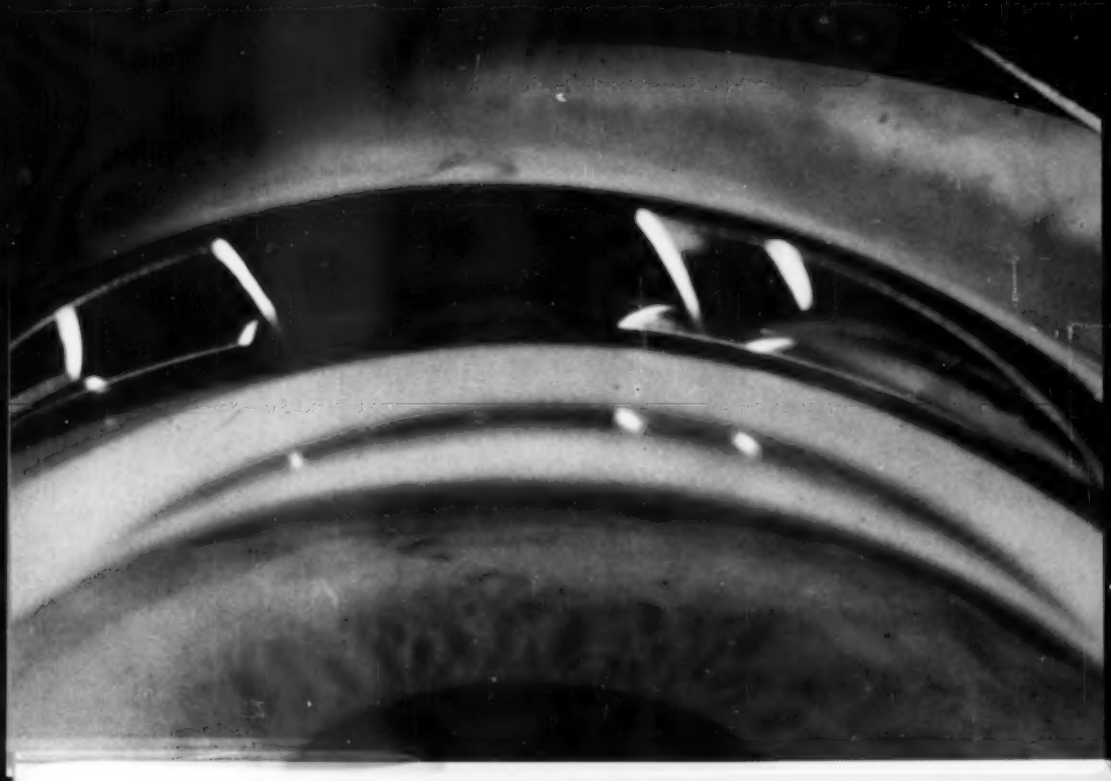

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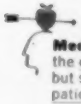

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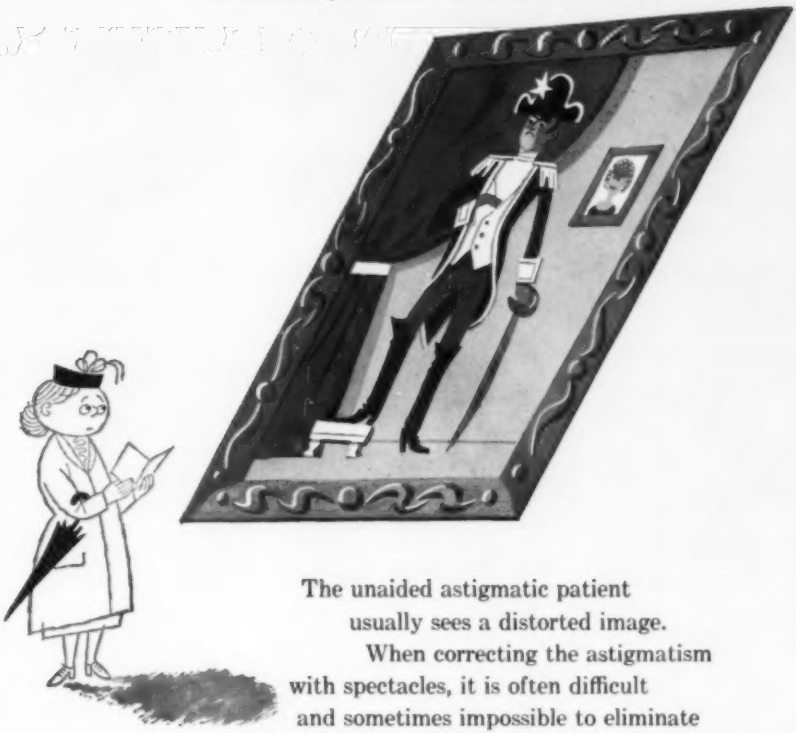

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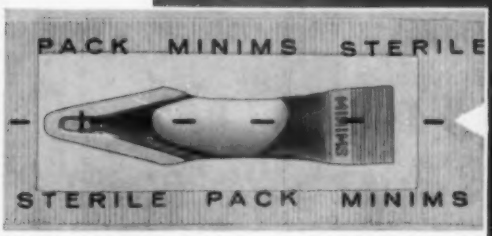

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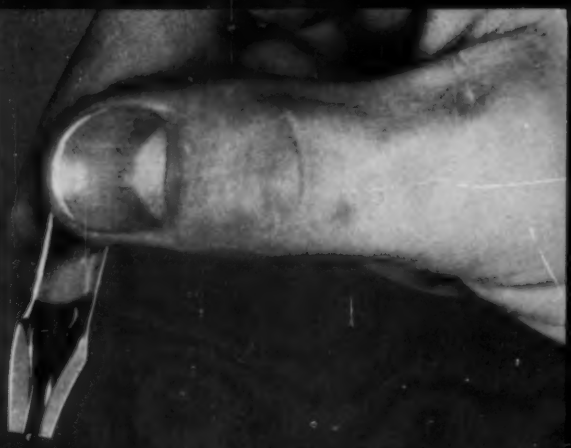

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Why is this important to presbyopes? Because the wearer no longer shows the telltale sign of aging: a visible bifocal segment.

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The new Younger-22 Seamless Lens is the first truly-invisible, optical-quality, dual-field lens. Not only is the segment invisible to the beholder; the wearer is not likely to be annoyed by the visible segment line; the soft transition zone between fields eliminates that.

Your presbyopic patients will welcome this ageless dual-field lens; convince yourself by trying a pair at your first opportunity!

The *New*
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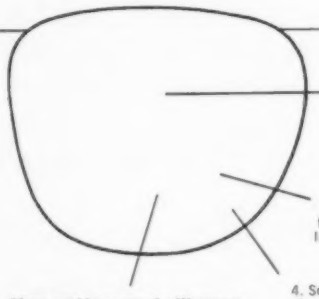
YOUR PATIENT TAKES NO RISK with Younger-22s. If the patient cannot wear them for any reason, it will be replaced with a fused bifocal of your choice at no additional cost.

Frame shown: Liberty's famous Broker 215



2. Front surface quality and spherical accuracy to millionths of an inch.

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WHEN AND HOW TO CORRECT VERTICAL IMBALANCE

We are all familiar with the characteristic complaints of a patient who has received bifocals, particularly first bifocals: without some form of compensation for vertical imbalance at the reading point—headaches after reading for only a short time, inability to fuse, suppression of one eye, etc. In order to avoid such complaints, we correct imbalances as low as 0.3 to 0.4 prism diopters. Whether correction for such slight amounts of vertical imbalance is necessary depends primarily on the use of the bifocal. You should correct this type of imbalance for an architect or an accountant but not for a salesman or housewife. In general, every imbalance over 1.00 prism diopter should be corrected. Perhaps because they are more frequent and more often overlooked, a surprising number of complaints arise from small uncorrected imbalances.

There are two instances when you should not correct the entire amount of induced vertical imbalance. First, when a bifocal wearer has accustomed himself to fusing with the imbalance; second, where a phoria exists which tends to balance the induced prism at the reading point.

THE FOUR METHODS OF CORRECTING VERTICAL IMBALANCE ARE:

1. DROP DISTANCE CENTERS This divides the imbalance between the distance and reading portion. Whatever it removes from the reading portion of the lens it adds to the distance. For this reason it is used in imbalances of less than 0.6 prism diopter.

2. DISSIMILAR SEGMENTS We recommend this procedure only when two bifocal segments of similar nature are used, such as a combination of Ultex A and E, flat top D and R, Kryptok 23mm and 20mm. How much imbalance may be corrected in this manner is a function of the bifocal addition. Its practical limits are in the range of 1.25 prism diopters.

3. PRISM SEGMENTS This is an extremely poor method because of the increased thickness of such a lens and its 1.50 prism diopter upper limit. It frequently is used for creation of horizontal prism in the segment only.

4. BALANCE CENTER This is the best method of correction for most imbalances. There is no upper limit but balance centers of less than 1.00 prism diopter are impractical to grind. Since it is always ground base up, the slab-off is used on the lens of the greatest minus power or the least plus power.

With some foresight in lens design, the ophthalmologist should never receive patient complaints about uncorrected or over-corrected vertical imbalance.

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AMERICAN JOURNAL OF OPHTHALMOLOGY

VOLUME 47 MARCH, 1959 NUMBER 3

THE PRESERVATION OF EYE TISSUES*

CURRENT STATUS IN TRANSPLANTATION

JOHN HARRY KING, JR.,† M.D., AND SHANKAR B. CHAVAN,‡ M.D.
Washington, D.C.

The use of tissues from a donor eye has become accepted as a valuable means for replacing and supplementing living ocular tissues in the restoration of vision, the prevention of blindness, and in the repair of damage caused by injury. Donor corneas stored in a refrigerator at 6°C. must be used within 72 hours. Pooled vitreous, however, has been implanted even after years of refrigerator storage. These materials are usually obtainable from eye banks for elective surgery, but for emergency use they are often unavailable to the average ophthalmologist.

The term "bank" is a misnomer when applied to the storage of eye tissues which must be used within short periods of time. Eye banks are actually "collecting stations" which distribute fresh tissues soon after they are obtained. Ideally, ocular tissues should be available at any time and at any place.

It is possible to preserve and store certain eye tissues indefinitely for use in various surgical procedures, although, at present, this is not being done generally by eye banks.

CORNEA

Corneas can be preserved and stored for prolonged periods and can be successfully used for lamellar keratoplasty. The results obtained with lamellar grafts preserved by two recently developed methods have shown that stored corneas can be used with success equal to that obtained when fresh donor tissues are employed. Eastcott and his coworkers[1] reported a method of preserving corneas by freezing after pretreatment with 15-percent glycerin. McNair and King[2] preserved animal corneas by dehydration in a mixture of 15-percent glycerin and isotonic saline and stored them at room temperature. King[3] extended these studies and dehydrated corneas in 95-percent glycerin, sealing the tissues in a vacuum and storing them at room temperature. Five human corneas, the oldest stored for two months, were successfully used for lamellar grafting. From the standpoint of economy and shipping, the preservation of corneas by dehydration in glycerin and storage at room temperature is obviously preferred to that of storage in the frozen state.

Corneas preserved by the method of King[3] have now been used in lamellar grafting in over 60 patients. The results are considered equal to those obtained with fresh donor tissues. The corneas used in these patients had been stored at room temperature for varying periods with the oldest cornea having been preserved for two years.

When a lamellar graft is required for therapeutic purposes in cases of acute or recalcitrant keratitis, the use of a preserved

* The corneal studies were supported in part by a research grant, B-975(R), from the National Institute of Neurological Diseases and Blindness of the National Institutes of Health, Public Health Service. These studies were also financed by the Washington Eye-Bank and Research Foundation (Lions 22-C), and the entire work on the vitreous, conjunctiva, sclera, and lens was supported by the Eye-Bank.

† Clinical Associate Professor of Ophthalmology, Georgetown University Medical School; Chairman, Eye Research, Washington Hospital Center.

‡ Research Fellow in Ophthalmology, Eye Research Section, Department of Surgery, Georgetown University Medical Center.

cornea appears to be superior to that of fresh tissue. The postoperative inflammatory reaction is less and the cornea clears more rapidly. Neovascularization is absent or less in the preserved graft but vessels may penetrate the host cornea.

As noted in the dehydration of arterial tissues,[4] some proteins must also be removed from the cornea during the dehydration process. Animal experimentation[5] has proved that this protein removal may result in less antigen-antibody reaction when preserved corneas are used. The trophic or clearing effect of the preserved graft on the surrounding diseased cornea is greater than that which follows the use of a fresh cornea. The therapeutic superiority of preserved material may be due, in part, to the concentration of antibiotics in the donor cornea when it is rehydrated in an antibiotic mixture* before transplanatation. The therapeutic effect can be enhanced by adding other medications to the rehydrating solution, such as the corticosteroid prednisolone which is of value in active interstitial keratitis. This medicament is, of course, contraindicated in the presence of herpetic keratitis.

The successful use in penetrating keratoplasty of corneas preserved by dehydration in glycerin has been reported by several surgeons.[6,7] Although these corneas can be advocated with impunity for lamellar grafting, there is not sufficient evidence available at this time to recommend their use in penetrating transplantation unless an emergency measure is required to save an eye.

If preserved tissues were held in reserve by eye banks and major hospitals, corneas could be made available to any eye surgeon for immediate use in emergency grafting in cases of injury, chemical burns, ruptured ulcers, and descemetoceles. A full-thickness penetrating graft for the improvement of vision is usually an elective procedure, and the operation can be scheduled for a time when a fresh donor eye is available.

* Neosporin solution, Burroughs Wellcome Co., Inc., Tuckahoe, New York.

Vitreous

Little use has been made in eye surgery of the grafting of donor vitreous. During the past 10 years several surgeons[8,9] have reported the replacement of cloudy or bloody vitreous by fresh vitreous with an occasional good result. Paufique and Moreau[10] state that the lack of success in vitreous transplanation seems to be due to several factors. First, the passage of fresh vitreous through a needle changes laminated vitreous gel to a fluid, an alteration which causes drastic physical and chemical changes. Secondly, even with the application of preplaced scleral sutures, it is always difficult to close the scleral opening after vitreous injection. To avoid these difficulties, the use of lyophilized or powdered vitreous, which reduces the volume of the gel but does not alter its chemical properties, has been advised.

Paufique and Moreau transplanted vitreous by placing vitreous powder into the eye through a special trocar injector inserted through the sclera. This was followed by the injection of 1.5 cc. of distilled water or saline solution. In aphakic eyes, the diseased vitreous was aspirated through the pupil with a syringe and powdered vitreous was introduced by means of a spud. One cubic centimeter of distilled water was then injected into the eye and a large air bubble was left in the anterior chamber. These investigators did not propose the lyophilization of vitreous primarily as a method of storage.

The demands for human vitreous have increased since Shafer[11] reported its successful use in implantation in treating complicated retinal detachments. He employed pooled fluid vitreous which had been stored by ordinary refrigeration at 4°C. Pischel,[11] although commenting upon the value of the vitreous implant, disagrees that such stored vitreous is "self-sterilizing." He reports having cultured various bacteria from several samples. The technique of vitreous implantation has not become widespread, mainly because of the unavailability of donor vitreous.

We have preserved vitreous by lyophilization and then sealed the powder in a vacuum in a tube. It can apparently be stored indefinitely at room temperature. In our studies, the vitreous was not pooled but was removed from individual human eyes within 72 hours after enucleation. A corneoscleral rim, including the iris diaphragm and lens, was removed and the hyaloid membrane, if it had not ruptured, was incised. Vitreous was aspirated through a 10-cc. syringe without a needle and two or three cc. of vitreous could thus be obtained. The vitreous was placed in a sterile Pyrex tube which contained either 30 minims of antibiotic solution* or 10-percent sodium sulamyd solution. The fluid was dehydrated by an apparatus similar to that used for the rapid freeze-drying of arterial segments. Lyophilized vitreous is reconstituted by an equal amount of normal saline or distilled water and resembles normal vitreous in consistency and transparency.

Lyophilized vitreous has been stored for as long as 12 months and it appears that it can be preserved indefinitely without changing its composition. Cultures taken on this material after varying periods of storage have shown no growth.

Preserved vitreous has been implanted in 16 patients with complicated retinal detachments and has been used as a vitreous transplant following injury or disease in five instances. It behaves in the same manner as fresh vitreous, clearing usually within the first week after operation and causing no adverse effects. No attempt was made to evaluate the efficacy of vitreous implant for the treatment of retinal detachment. It is felt, however, that preserved vitreous is as satisfactory for this purpose as is fresh vitreous.

Following the treatment of some complicated retinal detachments, we have implanted a "concentrated" preserved vitreous. The lyophilized powder obtained from 3.0 cc. of vitreous removed from an individual eye was reconstituted with 1.0 cc. of normal saline. The resulting thick solution was injected through an 18-gauge needle or the knife-needle of Amsler which was inserted through the choroid in a small scleral incision supported by a preplaced mattress suture. It was felt that this vitreous would undergo further hydration within the eye. A firm pressure of the eye was maintained and glaucoma was not observed in any patient.

Dried stored vitreous has the following advantages:

1. It is readily available and can be easily stored or shipped without refrigeration.

2. It appears to be more viscous after rehydration than pooled vitreous which has undergone prolonged refrigerator storage.

3. Either the powdered form or a concentrated form can be used which will undergo further rehydration within the eye. A more prolonged tampon or pressure effect is then obtained for the treatment of retinal detachment.

4. The inclusion of antibacterial agents is more dependable in assuring sterility than is pooled vitreous which is said to be "self-sterilizing."

5. Antibacterial agents added to donor vitreous may be beneficial to a diseased or injured eye after injection.

Conjunctiva

Autogenous conjunctiva is considered the best tissue for the replacement of defects of the conjunctiva resulting from trauma, symblepharon, removal of extensive pterygiums, or tumors. If enough conjunctival tissue needed to cover the defect cannot be obtained from the same patient for transplantation, other tissues may be employed. Mucous membrane taken from the lip or buccal area is preferred, although epidermal grafts are still used to replace an extensive defect, such as those which are necessary for the restoration of an obliterated socket. Several sugeons[12,13] have successfully used cadaver conjunctiva as a graft.

* Neosporin solution, Burroughs Wellcome Co., Inc., Tuckahoe, New York.

Although substitutes for conjunctiva may be satisfactory from a functional standpoint, in many instances their cosmetic result leaves much to be desired. The ideal tissue for the replacement of conjunctiva is donor conjunctiva. We have preserved conjunctiva by several techniques of dehydration and lyophilization and both methods appear to be satisfactory. The method of dehydration in 95-percent glycerin, with 30 minims of antibiotic solution added, makes the tissue soft and fragile. Conjunctiva preserved by lyophilization after pretreatment with 15-percent or 95-percent glycerin becomes hard and brittle. Preserved conjunctiva resembles normal conjunctiva after it is reconstituted with physiologic saline.

The bulbar conjunctiva is excised from the donor after removal of the eyeball. A suture is placed through the epithelial surface of the cadaver conjunctiva so that this surface may be identified later. The material is then rolled upon a plastic rod, placed in a sterile Pyrex test tube, and covered by 95-percent glycerin. Upon the completion of the dehydration or lyophilization process, the tube is sealed in a vacuum and stored at room temperature. When the tissue is needed, the tube is opened under sterile conditions. The tissue is then transferred to a shallow dish, covered with antibiotic mixture, and rehydrated for about 15 minutes before it is used. When preserved dehydrated homogenous donor conjunctiva or lyophilized conjunctiva previously treated by glycerin is transplanted in animals, it heals well and assumes the appearance of fresh autogenous conjunctiva. However, when conjunctiva preserved by lyophilization without pretreatment with glycerin was transplanted marked tissue reaction resulted. There was much edema and healing was slow.

Human conjunctiva has been preserved by dehydration in glycerin and is being stored, but as yet there has been no opportunity for its use in transplantation. Preserved conjunctiva could be used wherever fresh conjunctiva is needed—following injury with tissue loss and as a substitute for mucous membrane in severe chemical burns of the eye in applying the Denig treatment.

Sclera

It would be preferable to use cadaver sclera in many operations requiring the use of a tissue to reinforce or replace the sclera. Such conditions as ectasia, staphyloma, and progressive myopia can be repaired with sclera.[14–16] The availablity of donor sclera, however, poses a problem in furthering this type of surgery.

Sclera can be preserved and stored indefinitely at room temperature in vacuum-sealed tubes. We have utilized the method of dehydration in glycerin and also the technique of freeze-drying. Before the tissue is used, it is rehydrated for 15 minutes by immersing it in a saline antibiotic mixture.

We have transplanted preserved sclera in the eyes of rabbits, cats, and dogs after removing full-thickness discs and strips of sclera. The donor sclera was sutured in the defect with interrupted silk sutures and covered by the animals own conjunctiva or by a piece of preserved homogenous conjunctiva. The tissues healed normally and could not be distinguished from fresh transplants. Sclera dehydrated in glycerin becomes soft and fragile and must be handled carefully during transplantation. Sclera preserved by lyophilization after pretreatment with glycerin is less fragile. When the tissue is lyophilized without pretreatment in glycerin it becomes hard, takes longer to rehydrate, and healing is slower.

Lens

The successful transplantation of a human cadaver lens was reported by Čavka[17] in 1956. The lens was perfectly clear six months after transplanting and there was no elevation of the intraocular pressure at any time.

Hull[18] transplanted fresh kitten lenses in cats and all of the transplants became opaque.

We have experimentally transplanted fresh and preserved homogenous lenses in cats, rabbits, and dogs and most lenses turned opaque within a few days after operation. In one rabbit, the lens remained transparent for three weeks. The procedure is difficult to evaluate because of the technical handicap in removing animal lenses intracapsularly which have strong adherent zonules.

Attempts to preserve the fresh lens (human, monkey, horse, goat, dog, cat, and rabbit) by dehydration in glycerin and by freeze-drying have not been successful in our experiments. When animal lenses are immersed in fresh vitreous containing 20 minims of 95-percent glycerin and 30 minims of antibiotic solution and sealed in a tube in vacuum, they have been kept at 6°C. and have maintained transparency for as long as six months. Further studies are being done in an effort to preserve and transplant lenses in animals.

In conclusion, new developments in the preservation of eye tissues should greatly advance the progress of eye surgery. When the ideal eye bank becomes a reality, corneas, vitreous, conjunctiva, and sclera will be made available to any eye surgeon in any location at any time.

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THE EFFECT OF GLYCERINE, DEHYDRATION, MERTHIOLATE, AND ZEPHIRAN ON THE VIABILITY OF CORNEA*

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At the present time, most authorities agree that donor material for corneal grafting must be viable. In earlier studies,[1] we had shown that corneas which had been stored in a moist chamber at 4°C. remained viable up to three weeks, and that those which had been pretreated in 15-percent glycerine and saline, and frozen slowly to −79°C., remained viable for at least three months.

The present studies were undertaken to determine the effects of glycerine and antiseptics (in the concentrations and for periods used in eye-bank laboratories), and of dehydration, upon the viability of corneal tissues.

METHODS AND MATERIALS

The tissue culture methods used in these experiments were the same as those described in earlier reports.[1] Corneal tissues were obtained from freshly killed rabbits.

1. *Glycerine.* Corneas were stored in glycerine, varying in concentration from 10 to 100 percent, and in temperatures ranging from +56°C. to −79°C. Tissues which were to be frozen were allowed to sit for 60 minutes at room temperature in different percentages of glycerine. The eyes were removed from storage at intervals, thawed rapidly when frozen, and planted in tissue cultures. In all there were 1,194 tissues (58 of which had been stored at 56°C., 82 at 37°C., 318 at room temperature, 334 at 4°C., 50 at −20°C., 42 at −30°C., and 310 at −79°C.). Normal saline was used as a control for these experiments.

2. *Dehydration.* Ninety-one whole eyes and isolated cornea were treated in 100-percent commercial glycerine for one hour, and then dehydrated with a vacuum pump for 15 hours. The tissues were then stored in vacuum from two to 96 hours at room temperature. Forty-nine tissues were dehydrated by the same process and stored at room temperature in nitrogen gas from two to 17 hours.

3. *Antiseptics.* Fifty-six whole eyes and isolated corneas were allowed to stand at room temperature in merthiolate, and 108 were immersed in Zephiran. The solutions varied in concentration from 1:500 to 1:5,000, and for periods of from five to 60 minutes.

RESULTS

1. *Glycerine* (table 1). At −79°C., 75 percent of tissues pretreated in 15-percent glycerine and saline remained viable up to three months of storage, whereas only four percent of the corneas pretreated in 100-percent glycerine were viable after seven days. Storage in saline (used as a control) at this temperature resulted in only 12 percent of the tissues being viable after five hours.

At −20°C. 59 percent of the tissues pretreated in 15-percent glycerine and saline remained viable up to six days and at −30°C., 62 percent were viable up to three days. None of the tissues pretreated in 100-percent glycerine were viable.

At 4°C. 63 percent of the tissues stored in 15-percent glycerine and saline remained viable up to three weeks, but only 30 percent of the corneas pretreated in 100-percent glycerine were viable after 10 hours.

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TABLE 1

MAXIMUM GROWTH PERIOD OF CORNEAL CELLS FOLLOWING STORAGE IN DIFFERENT CONCENTRATIONS OF GLYCERINE AT VARIOUS TEMPERATURES

Percentage of Glycerine	Temperature (centigrade)	Number of Tissues Planted	Number of Tissues Growing		Maximum Growth Period
Saline Control	−79°	16	2	(12%)	5 hr.
	−30°	8	2	(25%)	1 hr.
	−20°	6	2	(33%)	1 hr.
	4°	67	26	(39%)	22 hr.
	RT	20	4	(20%)	48 hr.
	37°	18	12	(66%)	24 hr.
	56°	14	4	(28%)	5 min.
10	−79°	10	1	(10%)	24 hr.
	4°	12	10	(83%)	6 hr.
	RT	17	4	(24%)	24 hr.
15	−79°	32	24	(75%)	3 mo.
	−30°	24	15	(62%)	3 da.
	−20°	34	20	(59%)	6 da.
	4°	22	14	(63%)	3 wk.
	RT	23	8	(35%)	24 hr.
	37°	20	10	(50%)	7 hr.
	56°	12	4	(33%)	5 min.
20	−79°	12	1	(8%)	24 hr.
	4°	14	9	(64%)	48 hr.
	RT	19	1	(5%)	6 hr.
30	−79°	15	1	(7%)	24 hr.
	4°	18	5	(27%)	48 hr.
	RT	18	5	(28%)	16 hr.
40	−79°	16	0		0
	4°	21	4	(19%)	24 hr.
	RT	18	3	(16%)	48 hr.
50	−79°	44	2	(5%)	12 hr.
	4°	37	8	(22%)	24 hr.
	RT	39	10	(25%)	7 hr.
	37°	20	6	(30%)	3 hr.
	56°	12	4	(33%)	5 min.
60	−79°	16	0		0
	4°	22	3	(14%)	24 hr.
	RT	18	2	(11%)	2 hr.
70	−79°	16	0		0
	4°	17	3	(18%)	16 hr.
	RT	20	4	(20%)	6 hr.
80	−79°	45	2	(4%)	24 hr.
	4°	38	12	(32%)	7 hr.
	RT	39	8	(20%)	4 hr.
90	−79°	18	0		0
	4°	14	4	(29%)	6 hr.
	RT	17	8	(24%)	6 hr.
100	−79°	78	3	(4%)	1 wk.
	−30°	10	0		0
	−20°	10	0		0
	4°	52	16	(30%)	10 hr.
	RT	66	13	(20%)	10 hr.
	37°	24	2	(8%)	1 hr.
	56°	20	2	(10%)	10 min.

At room temperature, none of the tissues stored in glycerine, regardless of concentration, remained viable after a period 48 hours. Thirty-five percent stored in 15-percent glycerine and saline were viable up to 24 hours and only 20 percent stored in 100-percent glycerine were viable up to 10 hours.

At 37°C., 50 percent of the corneas of whole eyes stored in 15-percent glycerine and saline remained viable up to seven hours. In 100-percent glycerine, eight percent of the tissues were viable up to one hour when the whole eye was stored.

At 56°C., 33 percent of the corneas stored in 15-percent glycerine and saline remained viable up to five minutes. Only 10 percent of corneas of whole eyes pretreated in 100-percent glycerine remained viable up to 10 minutes.

2. *Dehydration.* Of the 140 tissues which had been dehydrated, none showed any evidence of viability.

3. *Antiseptics.* All tissues immersed for 10 minutes in a 1:5,000 solution of merthiolate remained viable, but, after 30 minutes, only 12 percent of excised corneas and 50 percent of corneas from whole eyes remained viable.

All tissues stored in Zephiran, in all concentrations, remained viable after immersion for one hour (at which point the experiment was discontinued).

Discussion

In these experiments, the protective effect of 15-percent glycerine and saline in the pretreatment of corneal tissues prior to storage at temperatures below the freezing point has been confirmed. In general, our findings were similar to those of Draheim and coworkers.[2] It is hard to explain why there was no growth following storage in concentrations of 40-, 60-, and 70-percent glycerine at −79°C., and also why there was such a small percentage of viable tissues following storage in 10 and 20 percent concentrations at −79°C., whereas maximum growth occurred when 15-percent glycerine was used. Fifteen-percent glycerine and saline could protect the corneal cells only when the temperature of storage was below freezing, but in any other concentration there was a marked decrease in viability (table 1). One hundred-percent glycerine does not appear to have any advantage in storage, at temperatures other than −79°C., where it provides slight protection.

Successful lamellar grafts, using dehydrated tissues stored in vacuum at room temperature, have been reported by King.[3] His criterion of viability was the postoperative transparency of the transplants. These tissue culture studies have shown that 100 percent of dehydrated tissues stored at room temperature either in vacuum or in an inert gas were nonviable. It seems possible, therefore, that viable tissues may not be necessary to produce a successful corneal graft.

Treatment of eyes in a solution of merthiolate 1:5,000, for a period of 10 minutes, is an accepted procedure in many eye-bank laboratories. In these experiments this did not affect the viability of the cornea. A solution of merthiolate stronger than 1:5,000 reduced the viability in isolated corneas. Zephiran in these experiments was less toxic to the corneal cells than merthiolate.

Summary

1. Whole eyes and isolated corneas were stored at various temperatures in varying concentrations of glycerine and antiseptics.

2. Other eyes and isolated corneas were dehydrated and stored in vacuum. All tissues were removed from storage at various intervals and the viability of epithelial cells and fibroblasts was determined in tissue cultures.

3. Glycerine, in concentrations of 15 percent, was found to have the greatest protective effect in the pretreatment of corneas prior to freezing.

4. In temperatures above freezing, glycerine in any concentration did not have any protective effect on the viability of the cells, and in a concentration of 100 percent appeared to have a toxic effect.

5. Dehydration and storage in vacuum resulted in nonviable tissues in 100 percent of specimens tested.

6. Merthiolate and Zephiran, in the concentrations and for periods used in eye-bank laboratories, did not affect the viability of corneal cells. After periods of immersion of 30 minutes or longer, Zephiran was less toxic than merthiolate.

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AN UNUSUAL TUMOR OF THE RETINAL PIGMENT EPITHELIUM*

In an eye with early open-angle glaucoma

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Tumors of the pigment epithelium of the retina may be divided into two groups. Comprising the first group are the discrete, sharply delineated black lesions in the fundus which are probably congenital in origin. Such lesions have been reported frequently in the literature under a variety of names—congenital pigmentary plaques of retina (Batten and Spicer[1]), congenital pigmentation of retina (Dodd[2]), grouped pigmentation of the fundus (Hoeg[6]).

Reese and Jones[7] have provided the most recent review of such lesions, terming them "benign melanomas of the retinal pigment epithelium." They placed the changes in the pigment epithelium "because of the black color, the sharp outline, and the clarity of the granular tissue markings." None of the lesions in their series was observed to increase in size and field defects corresponding to the lesions were not elicited. They were able to study one of these lesions microscopically and found that the pigment epithelium was reduplicated. In the choroid beneath this lesion, the melanoblasts were more pigmented. In their opinion, this lesion represented a "localized, congenital overgrowth of pigment epithelium."

In the second category are the group of cases reviewed by Greer,[5] in 1952, and more recently by Fair,[3] in 1958. In this group of cases, the lesion was characterized by an exuberant proliferation of retinal pigment epithelium. The tumor cells, large polygonal in shape and frequently nonpigmented, tended to arrange themselves in sheets, cords, or tubular masses. Invasion of the choroid, retina, or optic nerve was not unusual.

The majority of the tumors in this category have been found in eyes which have been the site of a prolonged inflammation. Thus, in each of the three cases accepted by Greer[5] as authentic, it was considered that the inflammatory lesion had provided the initial stimulus for hyperplasia of the pigment epithelium, only to be followed by a massive overgrowth of the epithelial cells to form a neoplasm.

Prolonged intraocular inflammation is not, however, a necessary prerequisite for the development of these tumors. In the cases reported by Rønne,[8] and by Theobald and her group,[9] there was neither clinical nor histopathologic evidence of ocular inflammation, the eyes having been removed for suspected malignancy.

In the case to be reported here the problem of clinical evaluation of the pigmented lesion in the fundus was further complicated by the development of retinal detachment.

Case report

The patient, a 39-year-old white man (JHH

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#756681), was first seen at the Wilmer Institute on March 6, 1957, with the complaint of poor vision in the right eye. He had one brother who was under treatment elsewhere for pigmentary glaucoma.

The patient's ocular difficulties had begun one month earlier, in February, with blurred vision in the right eye. Examination by an ophthalmologist in another city revealed a best corrected vision in the right eye of 20/70, edema of the macular area, and visual field changes consisting of an enlarged blindspot and a questionable central scotoma. As there was also a sphenoidal sinusitis on the right, two irrigations of the sphenoid sinus were performed, followed eventually by right sphenoidectomy. On March 1, 1957, the vision had improved to 20/30.

At the time of the initial examination at the Wilmer Institute, vision in the right eye could be corrected to 20/15. However, after ophthalmoscopic examination when the vision in the left eye had recovered to 20/25, the vision in the right eye was still reduced to 20/40. Patching of the eye for two hours resulted in improvement to 20/25. On ophthalmoscopic examination, the right fovea appeared more orange than the left but no definite macula edema was noted. There was also a nuclear and an anterior subcapsular opacity in the upper nasal quadrant of the right lens. The appearance of the macula plus the observed decrease in visual acuity following exposure to the examiner's lights suggested the diagnosis of early macular degeneration in the right eye.

Re-examination on July 16, 1957, revealed a corrected visual acuity of 20/20, although there had been an increase in the strength of the myopic correcting lens. However, slitlamp examination disclosed fairly dense nuclear sclerosis with some increase in the upper nasal quadrantic cortical lenticular opacity.

On ophthalmoscopic examination, a lesion in the midperiphery of the superior nasal quadrant of the fundus was observed for the first time. This was a jet black pigmented lesion about four disc diameters in width. Retinal vessels could be traced to the margin of the lesion where they became enveloped in pigmented retina (fig. 1).

Examination of the margin of the lesion with the Hruby lens and slitlamp showed no elevation of the retina at this point. The center of the lesion was so black that a satisfactory reference point could not be obtained. For this reason various observers, even when using the Schepens indirect binocular ophthalmoscope, described the lesion as flat or as elevated up to six diopters. All, however, agreed that the retina over the lesion was not detached. With the Goldmann perimeter, an absolute scotoma corresponding to the size of the pigmented lesion was demonstrated (fig. 2).

Several diagnoses were entertained—malignant melanoma of the choroid, benign nevus of the choroid, choroidal hemorrhage. As the lesion was relatively small and could be easily observed, and since an exact scotoma corresponding to the tumor could

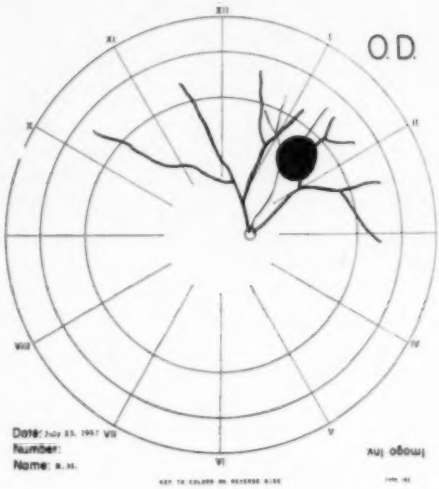

Fig. 1 (Duke and Maumenee). Drawing of pigmented lesion in superior nasal quadrant of right eye, as seen with Schepens indirect ophthalmoscope.

be outlined, it was decided to follow the patient closely.

On August 21, 1957, the ocular tension of the right eye was found for the first time to be elevated to 35 mm. Hg (Schiøtz); that in the left eye was 23 mm. Hg (Schiøtz). Gonioscopic findings in the two eyes were similar—an open angle throughout, a considerable deposition of pigment in the trabecular meshwork (grade 2), and on the posterior surface of the lens capsule near the equator, a glistening ring of orange-brown pigment.

On September 4, 1957, further diagnostic studies pertaining to the glaucoma were performed. In the right eye the ocular tension was 22.4 mm Hg (Schiøtz) and facility of outflow was 0.15; after the water-drinking test the ocular tension was 34.0 mm. Hg, and facility of outflow was 0.05. In the left eye the ocular tension was 17.0 mm. Hg (Schiøtz) and the facility of outflow was 0.05; after the water-drinking test the ocular tension was 31.0 mm. Hg (Schiøtz) and the facility of outflow was 0.04.

At biweekly intervals between July 16 and September 4, 1957, the patient was examined thoroughly, including visual field studies with the Goldmann perimeter. On September 4, 1957, the pigmented fundus lesion and the corresponding scotoma in the right eye were unchanged. However, 10 days later, the patient began to note photopsia and a veil-like encroachment upon the temporal field of vision of his right eye. On examination five days after the onset of these symptoms, visual acuity, right eye, was reduced to 3/200 in the upper field. On ophthalmoscopic examination, there was a billowing detachment of the retina above, extending from the

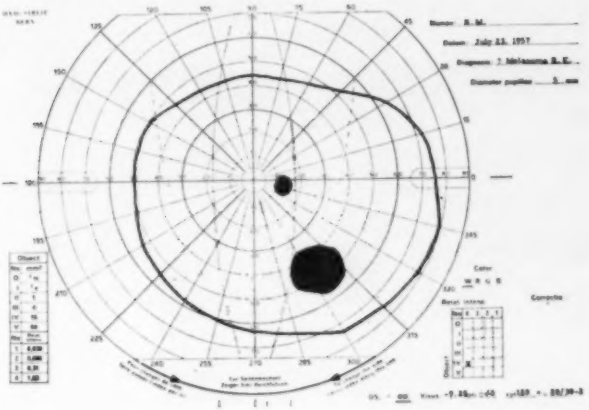

Fig. 2 (Duke and Maumenee). Scotoma in lower temporal field, right eye, as charted with Goldmann perimeter.

8:30- to the 2:00-o'clock positions. A hole could not be found in the retina. There was an additional unusual finding. Part of the pigmented lesion appeared to have been detached with the retina, for, on examination with the Hruby lens and slitlamp, the pigment was noted lying within the detached retina itself.

This observation cast some doubt on the diagnosis of malignant melanoma of the choroid, as it was considered unusual for a choroidal melanoma of this small size to show this degree of extension into the retina. However, because five consultants had made the diagnosis of malignant melanoma and the retinal detachment could not be explained, the eye was enucleated on the following day.

Gross examination. When the eye was sectioned, clear fluid gushed out. Some of this had been beneath the retina but on completion of the section, the retina fell nicely back into place. In the upper nasal quadrant, just posterior to the equator was a flat, round, black lesion seven mm. in diameter (fig. 3). The retina over this lesion had a central area of dark-brown pigmentation about four mm. in diameter (fig. 4). Grossly, no changes were observed in the macula region. No hole was found in the retina. The vitreous was partially liquid, formed vitreous remaining anteriorly.

Microscopic examination. The anterior chamber was of normal depth and the chamber angle was open. Within the spaces of the trabecular meshwork a small amount of brown melanin pigment was noted, though this pigment deposition was not as striking on microscopic examination as it had been clinically. There was no thickening of the fibers of the trabecular meshwork and the canal of Schlemm

Fig. 3 (Duke and Maumenee). The large pigmented tumor mass can be seen lying at the equator to the left of the optic disc. (Reproduced from Kodachrome transparency.)

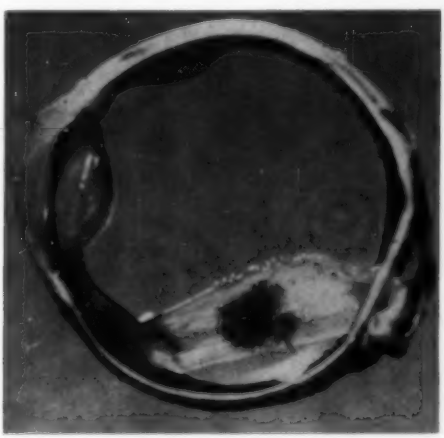

Fig. 4 (Duke and Maumenee). The pigmented lesion within the retina viewed from the outer surface of the artifactually detached retina. (Reproduced from Kodachrome transparency.)

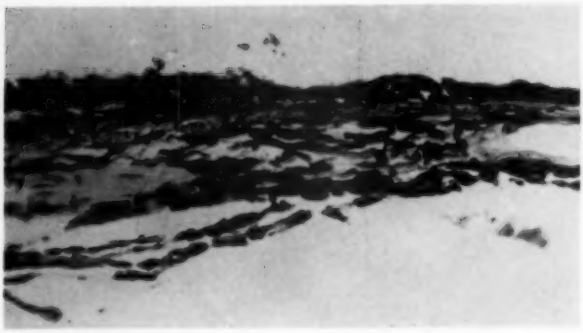

Fig. 5 (Duke and Maumenee). Zone of transition between normal retinal pigment epithelium on the right and the thickened pigment epithelium comprising the tumor on the left. (Hematoxylin and eosin; ×220.)

was widely patent. Serial sections revealed no sclerosis of the collecting channels or of the vessels of the deep scleral plexus. The iris was essentially normal, though there was fairly heavy pigmentation of the anterior limiting layer.

In the retina, located at the equator and extending a short distance posterior to it, there was a rather striking localized lesion. Here, the pigment epithelium was reduplicated upon itself, being two or three cell layers in thickness. At each extremity of the lesion, the transition between normal and abnormal pigment epithelium was abrupt (fig. 5). The inner layer of this reduplicated pigment epithelium was separated from the outer layer by a thin eosinophilic line, which suggested the development of Bruch's membrane (fig. 6). Beneath the outermost layer of pigment epithelium, the true Bruch's membrane could be seen in its normal location. The cells comprising the lesion were larger than normal, and were completely filled with the rod-shaped granules of melanin pigment, characteristic of that of pigment epithelium. Depigmented sections revealed that the tumor was composed of large cuboidal cells with rounded nuclei situated at the base of these cells. No mitotic figures were noted.

There was no extension of the tumor into the choroid, nor was there increase in choroidal melanoblasts. There was no significant degree of inflammatory reaction in the choroid.

In the region of the retina which lay directly over the lesion of the pigment epithelium, there was almost complete disruption of the normal retinal architecture. Rods and cones were entirely absent and the components of the nuclear and ganglion-cell layers were markedly reduced in number, with disorganization of those elements remaining. Numerous, heavily pigmented cells from the tumor of the pigment epithelium diffusely infiltrated the retina at this site (fig. 7). Most numerous at the outer aspect of the disorganized retina, a few of the pigmented cells extended inward to the nerve-fiber layer (fig. 8). Peripheral to this retinal lesion, there was degeneration of the rods and cones in varying degree and toward the ora serrata these elements disappeared entirely. Throughout the entire posterior pole of the retina, and especially affecting the macula region, there was extensive cystoid degeneration of the outer plexiform layer of the retina, the spaces in some instances being filled with a homogeneous eosinophilic material (fig. 9). The rods and cones, however, were here well preserved.

In the foveal region, there was some loss of the nuclei of the outer nuclear layer, resulting in considerable thinning in this region, though no foveal hole was found. The extent of the retinal detachment could not be accurately evaluated because most of the subretinal fluid had escaped when the eye was sectioned grossly.

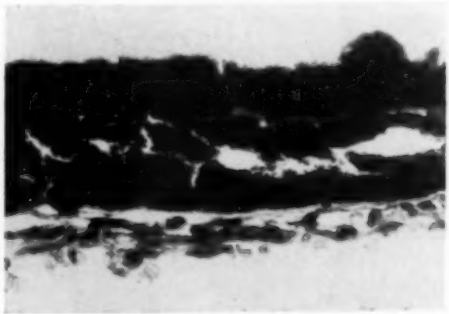

Fig. 6 (Duke and Maumenee). Section through center of the tumor. The newly formed Bruch's membrane can be seen as a thin horizontal line in center of pigmented lesion. True Bruch's membrane and choriocapillaris are at base of the pigmented lesion. (Hematoxylin and eosin; ×650.)

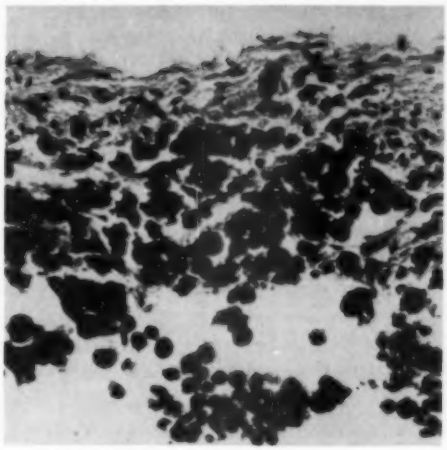

Fig. 7 (Duke and Maumenee). Section through area of retina which overlay the tumor, showing disorganization and infiltration of retina by pigmented tumor cells. (Hematoxylin and eosin; ×220.)

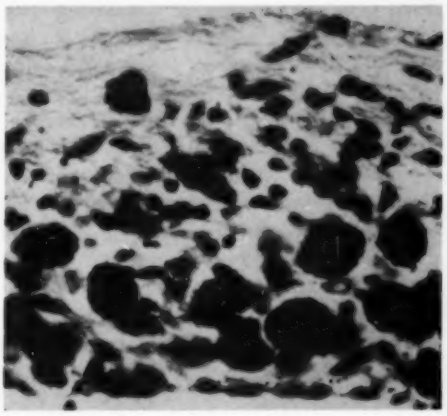

Fig. 8 (Duke and Maumenee). High-power view showing infiltration of retina by large pigmented tumor cells. The inner surface of retina is above. (Hematoxylin and eosin; ×650.)

Discussion

Despite the fact that the pigmented fundus lesion was not observed at the time of initial examination, it is quite probable that it was present at that time. In fact, there are many features which suggest that the lesion falls into the group described by Reese as congenital, benign melanomas of the pigment epithelium of the retina. The lesion was flat, sharply demarcated, heavily pigmented, and showed no increase in size during two months of observation. However, in the present case, the retina over the lesion was infiltrated by the tumor cells and there was a

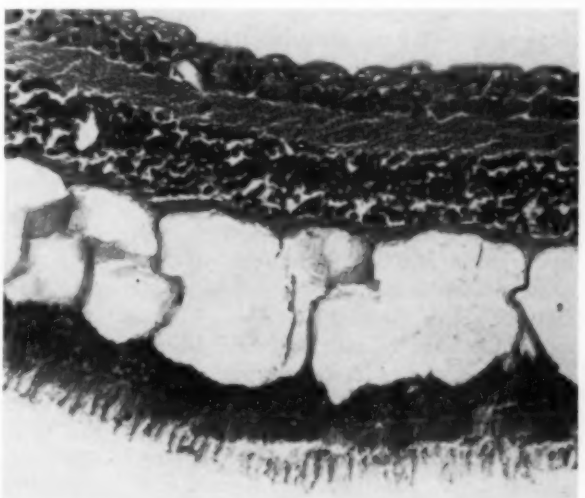

Fig. 9 (Duke and Maumenee). Extensive cystoid degeneration of outer plexiform layer of retina at posterior pole of the eye. (Hematoxylin and eosin; ×220.)

corresponding defect in the visual field—findings which were not noted in Reese's group of cases. The invasion of the retina might suggest that the lesion belongs to the group of actively proliferating tumors described by Fair,[3] for, previously noted, invasion of contiguous structures—retina, choroid, optic nerve—was not unusual in that group of cases. However, in each of the cases in that category, the tumor presented features of a more varied and active cytologic pattern —the arrangement of cells in sheets and cords, variability in degree of pigmentation, and the presence of mitotic figures. In the tumor under discussion here, on the other hand, the cytologic pattern was quite uniform, and no mitotic figures were found at all. Only the rather extensive invasion of the retina overlying the tumor suggested any degree of activity within the lesion. One had perhaps best place this lesion somewhere between the pigmented, quiescent, congenital tumors and the actively proliferating, invasive tumors.

Features other than the problem of classification are obscure as well. The extensive cystoid degeneration of the posterior pole of the retina that was found on microscopic examination may have been related to the macula edema noted by the first examiner and suspected but not unequivocably present at subsequent examinations. It is unlikely that the retinal degenerative changes could have been related to the extensive retinal detachment, as that condition had been present for only one week prior to enucleation.

One recalls the two cases of choroidal melanoma in the temporal periphery cited by Fry and McDonald.[4] In each of these cases, the initial symptoms were related to a disturbance in the macula region. In one case, the macula, clinically, had the characteristic appearance of central serous retinopathy, and in the other, histologic examination revealed serous detachment of the macula with small cystic spaces in the inner nuclear layer. Klien, in her discussion of these cases, cited an eye with malignant melanoma of the iris which on histologic examination showed a typical serous retinopathy in the macula region. Some type of vascular damage by toxic by-products of the tumor was considered as a possible cause for the macula disturbance.

It is worthwhile to emphasize that in the case under consideration here the macula edema was associated with a tumor of the pigment epithelium of the retina, probably a neoplasm of benign type, whereas in the previous cases cited in the literature, the macula disturbance had been associated with a malignant tumor of the uveal tract.

The mechanism of the retinal detachment also remains obscure. The tumor of the pigment epithelium was not sufficiently elevated to cause detachment and no retinal holes or traction bands were found.

The patient was thought by one observer to have pigmentary glaucoma. Clinically, there was a Grade 2 deposition of pigment in the trabecular meshwork and while the patient was under observation, elevation of the ocular tension in both eyes developed. Each eye after the water-drinking test showed a significant increase in the ocular tension and a drop in the facility of outflow. In the eye examined histologically, the chamber angle was open and there was little pigment in the trabecula. However, Dr. Lorenz Zimmerman of the Armed Forces Institute of Pathology performed alcian blue stains on sections of this eye and was unable to demonstrate stainable acid mucopolysaccharide in the intertrabecular spaces, though the mucopolysaccharides in other portions of the eye did take the stain. The tension in the remaining eye has been easily controlled with pilocarpine for six months.

Summary

A case of a tumor of the pigment epithelium of the retina with a visual field defect has been described. The patient initially sought medical attention because of decreased vision associated with macula edema. The development of retinal detachment pred-

icated removal of the eye in order to rule out the possibility of choroidal malignant melanoma.

On histologic examination the tumor of the pigment epithelium had invaded the retina. In the macula and perimacula regions there was extensive cystoid degeneration of the retina.

While under observation, the patient developed glaucoma associated with a wide chamber angle and characterized by a significant elevation of ocular tension and a significant decrease in facility of outflow, following the water-drinking test. The eye was enucleated one month after the first recorded elevation of ocular tension and histologic examination revealed an essentially normal trabecula, a widely patent canal of Schlemm, no sclerosis of the collecting channels, and absence of stainable acid mucopolysaccharide in the intertrabecular spaces.

The Johns Hopkins Hospital (5).

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CLINICS IN PERIMETRY NO. 3*

Incongruous homonymous hemianopsia

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Lesions that interrupt the optic tracts between the chiasm and the lateral geniculate bodies produce homonymous defects in the visual fields that are quite dissimilar, or incongruous. Defects of this nature are rare. Some are caused by pituitary tumors which, instead of growing upward, have extended backward. Adamantinomas, too, may involve the tracts, as may aneurysms of the circle of Willis. Other lesions of the optic tracts, in our experience, are even rarer and include glioma and multiple sclerosis. A few examples, selected somewhat at random, will supply the basis for this paper.

Aneurysm of the circle of Willis

Case 1

A 46-year-old woman was seen at the Mayo Clinic on June 18, 1951. Her only complaint was blurred vision of about six months' duration. She had noted that while driving a car she could not see as well to the right as to the left. Her visual fields had been plotted and a diagnosis of a toxic amblyopia, possibly due to tobacco, had been en-

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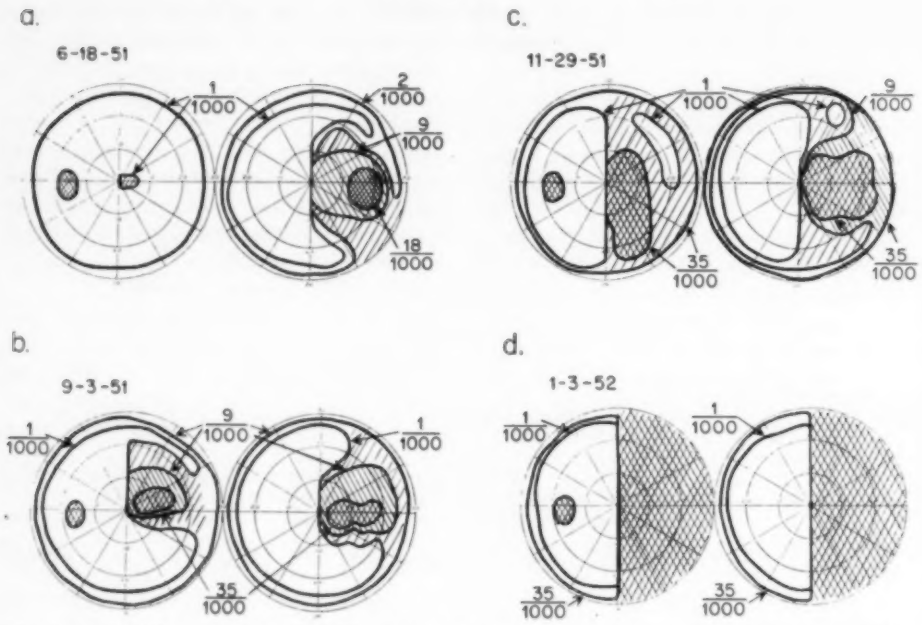

Fig. 1a to 1d (Kearns and Rucker). Progression of incongruous homonymous hemianopsia caused by aneurysm of circle of Willis.

tertained. A neurosurgeon had suggested a pneumoencephalogram, but this had been deferred.

On examination we found the ocular fundi to be normal. Incongruous right homonymous scotomas were demonstrated on the tangent screen (fig. 1a). These fields were interpreted as being indicative of a lesion of the left optic tract. General and neurologic examinations revealed no other abnormalities, and the results of the laboratory examinations including roentgenogram of the head were normal. Neurosurgical consultation was obtained, and a course of X-ray therapy to the left parasellar region was recommended and given. The patient was asked to return for re-evaluation and possible pneumoencephalographic study or exploration.

She returned on September 3, 1951, and, as can be seen in Figure 1b, the scotomas had progressed considerably. On September 6th, air encephalography was carried out. This demonstrated that the ventricles filled and were of normal size and shape but there was a filling difficulty deep on the left side near the third ventricle which suggested the possibility of a small space-occupying lesion in this region. Another course of X-ray therapy was given, and the patient was asked to return in three months.

On November 29, 1951, the scotomas were found to have enlarged even further (fig. 1c). On December 4th, a left carotid angiogram revealed an aneurysm about one inch in diameter arising from the carotid artery proximal to its bifurcation. On December 10th, the left internal carotid artery was ligated in the neck and nine days later the region of the aneurysm was explored through a left transfrontal craniotomy. The aneurysm was visualized and was found to be compressing the optic chiasm. The internal carotid artery was ligated close to the aneurysm. Following this operation, the patient was aphasic, but slowly improved over the succeeding weeks. On January 3, 1952, the visual fields were replotted, and complete right homonymous hemianopsia was found (fig. 1d). This was interpreted as the result of infarction of the left optic radiation superimposed on the previous field defects.

Astrocytoma

Case 2

On July 16, 1956, a 17-year-old girl registered at the clinic with a complaint of having had headache and stiff neck for two months. Four days prior to admission she had experienced a severe headache which she described as feeling that her head was going to explode. Nausea and vomiting were associated with this severe headache. A physician in her home locality had found that she had choked discs and referred her for neurosurgical consultation.

On examination in the eye section of this clinic,

vision was found to be 20/20 in each eye. The optic nerveheads were choked, the right was elevated six diopters, the left, five diopters. The discs were somewhat pale, although the veins were engorged and there were a few small retinal hemorrhages and exudates. Plotting of the visual fields showed an incongruous right homonymous hemianopsia (fig. 2a) indicating a lesion involving the left optic tract.

The neurologist stated that the patient probably had a cerebellar lesion because of some tremor of the right hand and stiffness of the neck. Roentgenograms of the head showed enlargement of the sella turcica with destruction of the posterior clinoids and the anterior clinoid on the right. There was also marked accentuation of the convolutional markings by increased intracranial pressure.

On July 18, 1956, a ventriculogram was performed. Marked hydrocephalus of the lateral ventricles was demonstrated, but the third and fourth ventricles did not fill. The contour of the lateral ventricle suggested the presence of a tumor in the region of the third ventricle. Left transfrontal craniotomy was carried out, and after the dura was incised along the wing of the sphenoid, a tumor was found arising from the side of the optic chiasm. Biopsy was done, and the pathologist reported the lesion to be a grade 1 astrocytoma of the chiasm.

On July 24th, the fields were again plotted and showed additional loss of field (fig. 2b).

Pituitary tumor

Case 3

A 34-year-old woman was seen at the clinic in May, 1955, at which time a diagnosis of Cushing's disease was made. Bilateral adrenalectomy was performed at that time. Since a number of patients with Cushing's disease are found to have a chromophobe adenoma of the pituitary gland, a roentgenogram of the head was obtained. The sella appeared generous in size but otherwise normal. The patient had no visual symptoms, and, although

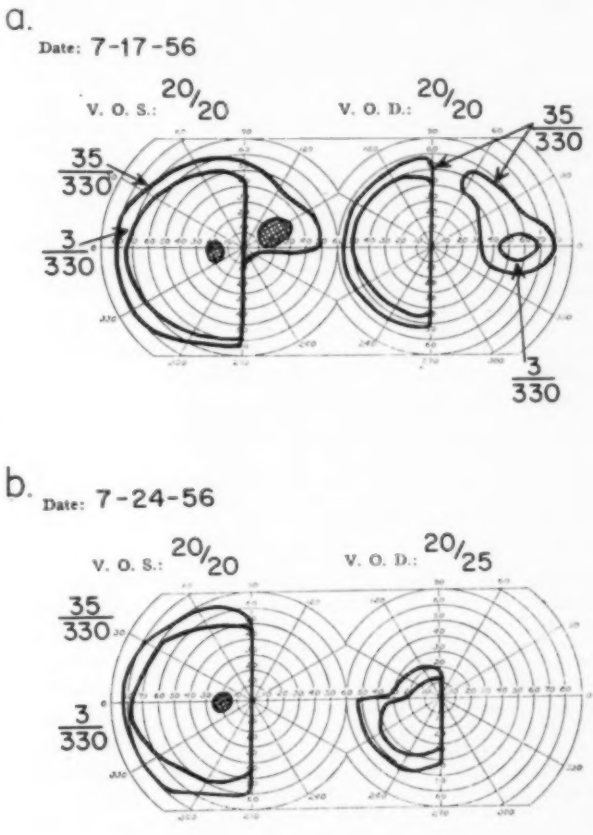

Fig. 2 (Kearns and Rucker). Incongruous homonymous field defects caused by astrocytoma of the chiasm. (a) Before operation. (b) After operation.

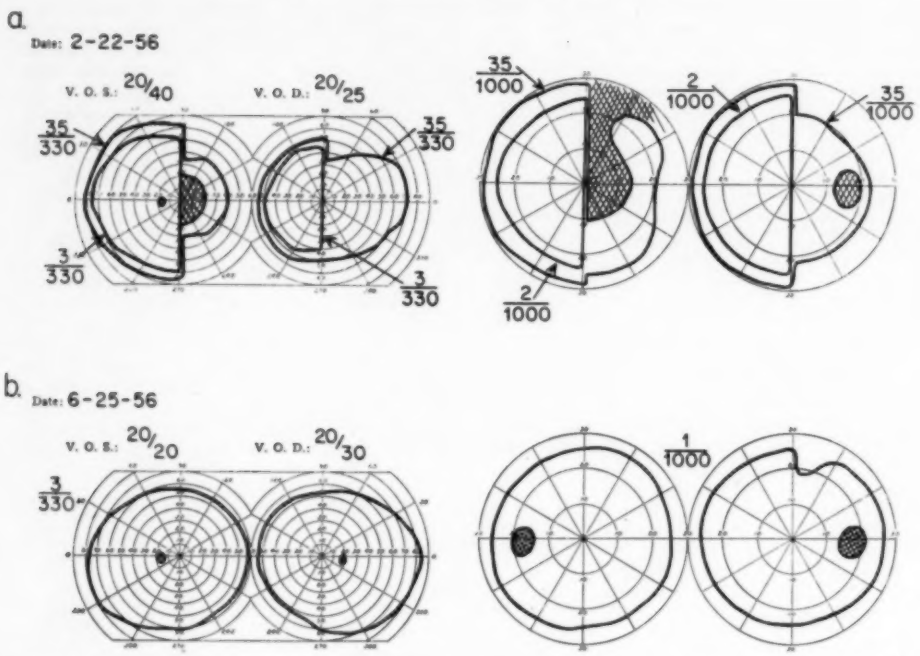

Fig. 3 (Kearns and Rucker). Incongruous right homonymous hemianopsia produced by a chromophobe adenoma of the pituitary gland. (a) Before operation. (b) After operation.

the fields were not plotted, the gross fields were normal. Ophthalmoscopic examination revealed only moderate hypertensive arteriolar sclerosis.

She returned to the clinic in February, 1956, because of recent onset of blurred vision. Partial paralysis of the left third nerve was present as evidenced by abnormal ocular rotations, a dilated left pupil, and ptosis of the left upper lid. The visual fields were plotted and an incongruous right homonymous defect with some loss of central vision on the left was found (fig. 3a). The fundi were normal except for the previously described hypertensive changes. The ocular findings were interpreted by the ophthalmologist as being indicative of a left parasellar lesion. On the basis of previous similar cases he predicted that the lesion was most likely a chromophobe adenoma growing laterally out of the sella. The roentgenogram of the head showed "decalcification of the right side of the dorsum sella and the right anterior clinoid."

An angiogram was done on February 25, 1956, and the left internal carotid artery appeared distorted by a parasellar mass. Three days later left frontoparietal craniotomy was done and a bilobulated mass was found extending to the left of the sella. As much of the tumor mass as possible was removed, and the pathologist reported it to be a cellular chromophobe adenoma.

On March 2nd, the fields showed considerable improvement, and the third nerve palsy was also less marked. The patient returned to the clinic for reexamination on June 25, 1956, at which time the fields were essentially normal (fig. 3b). The only evidence of third nerve palsy was slight weakness of upward rotation of the left eye.

Comment

Each of these three patients had incongruous homonymous defects. This marked incongruity is characteristic of lesions of the optic tract and is explained by the fact that the corresponding nerve fibers of each eye are more separated in this area than they are farther back in the optic radiations. Slight incongruity does occur in lesions of the anterior optic radiations but never to the extent shown in these examples.

The same lesions that give rise to bitemporal hemianopsia may at times produce incongruous homonymous defects. Although they are then referred to as tract lesions, they are essentially chiasmal lesions that in-

stead of expanding in the midline have involved the optic tracts of one side.

Cerebrovascular accidents do not produce bitemporal hemianopsia nor markedly incongruous homonymous defects. The perimetrist may state with certainty where the lesion is located but on the basis of the visual fields alone he is not able to state what the lesion is.

SUMMARY

Incongruous homonymous hemianopsia is an unusual type of visual field defect. It is indicative of a lesion of the optic tract and may result from the same types of lesions that give rise to bitemporal hemianopsia. The case histories and visual fields of three such cases are presented.

Mayo Clinic.

INCONTINENTIA PIGMENTI

ASSOCIATED WITH CHANGES IN THE POSTERIOR CHAMBER OF THE EYE

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New York

A boy, aged four and one-half months, recently came to us for consultation because of abnormalities of both eyes, which on examination proved to involve a mass in the posterior chamber. Because of a striking skin condition, a dermatologist was consulted and on the basis of clinical examination and histologic study, the case was diagnosed as one of incontinentia pigmenti. Recent reviews of this rare skin disease suggest that about one third of all cases have associated eye anomalies, and that perhaps one third of the latter involve masses in the posterior chamber. Since these masses present certain rather difficult problems of diagnosis, and consequently of management, we are presenting a study of the few cases previously reported in the literature, adding our own, as a contribution toward clarifying understanding of this rare and rather confusing ocular anomaly.

INCONTINENTIA PIGMENTI SYNDROME

The skin condition appears at birth, occasionally as late as two years after birth, and with few exceptions only in girls. While it is occasionally associated with premature birth, most infants are born at full-term. While genetic factors seem clearly implicated in a few instances and, in a few others, infections or other abnormalities have been present during gestation, none of these seems to be present in most instances. Our own patient was a boy, and we found no evidence of genetic factors or of abnormalities during gestation.

The typical skin lesion consists of rather bizarre patterns of blue-gray to chocolate-brown splashes, flecks, or streaks arranged according to Blaschko's nevus lines. They affect the torso and extremities but not the face. Characteristically the pigmentation is preceded by an erythematous, vesicular, and bullous dermatitis, which is succeeded by a stage resembling linear verrucous nevi, which in turn fades out to be replaced by the typical pigmentation.[1] In some cases the first stage tends to disappear and recur, in which instances all stages may be irregularly present at the same time. At times the first stage follows the typical pattern of distribution sufficiently to make diagnosis possible, but in other instances it may be difficult to differentiate from such other bullous dermatoses as contact dermatitis, bullous impetigo, or epidermolysis bullosa. The pigmentation itself usually fades out during early childhood but may persist longer, even occasionally leaving traces in adulthood. No known treatment essentially or permanently affects this course.

Histologic examination during the stage of pigmentation reveals typically (1) some

signs of degeneration in the basal layers of the epithelium, with some irregular decrease in the amount of melanin; (2) an increase in the number of chromatophores bearing melanin in the upper parts of the corium. Bloch in 1926 considered that the disease was due to abnormalities of the basal cells, which were "incontinent" of melanin, permitting it to drop into the corium instead of eliminating it upward normally, and devised the name "incontinentia pigmenti" in accordance with this concept.[2,3]

Siemens,[4] in 1929, noticed the coincidence of other anomalies with the skin condition and Sulzberger,[5] in 1938, stated that the relationship was definite. A survey of current literature suggests that the consensus today is that incontinentia pigmenti is one manifestation of a syndrome of dysplasias of ectodermal and mesodermal tissues. The associated anomalies may be cutaneous, dental, musculoskeletal, cerebrospinal, ocular; any may occur as a sole associated defect, or several may be present in some cases, and apparently may occur in any combination. The exact incidence of such defects is difficult to ascertain since some reports are incomplete, but Carney[1] found them reported in 22 of 37 cases and discovered evidence that there were none in only five of this group.

Eye anomalies have been summarized by several authors.[1,6] Lieb and Guerry[7] found five cases of strabismus, one of nystagmus, two of blue sclera, one of myopia, five of cataract, one of papillitis, one of exudative chorioretinitis, three of retrolental fibroplasia, three of pseudoglioma, four of optic nerve atrophy, and two of metastatic ophthalmia. At least the simpler of these defects are known to occur congenitally at times, and present no particular problems of diagnosis. They need to be noted here because (1) they may occur singly or in combination as part of the incontinentia pigmenti syndrome; (2) they may occur in any combination with the associated dysplasias most readily classed as masses in the posterior chamber.

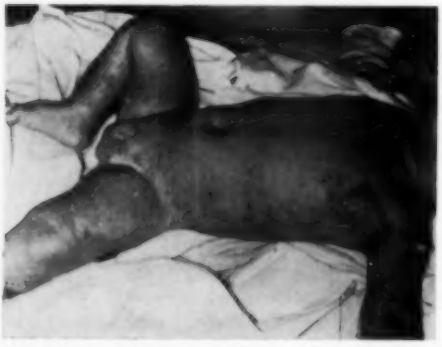

Fig. 1 (Cole and Cole). Distribution of pigment in the skin.

Mass in the posterior chamber

Seven cases of typical incontinentia pigmenti syndrome have been found in the literature with definite mass in the posterior chamber, with our own case making a total of eight. In all instances the diagnosis was verified by histologic study of the skin lesion. The presence of other anomalies, of genetic factors, of abnormalities during gestation, and sex were typical of other cases without the mass.

1. Bloch,[2] 1926: The right eye was enucleated during the first month of life with a diagnosis of glioma. There is no report of the left eye, and no pathologic report on the enucleated eye.

2. Haxthausen[8] and Hielesen,[9] 1945 and 1948: At about the eighth month the right eye was enucleated for pseudoglioma. The typical skin lesion appeared one month after operation. There is no report of the left eye and no pathologic report on the enucleated eye.

3. Uebel,[10] 1950: In both eyes ptosis palpebrarum, strabismus convergens, and deorsum vergens were observed. In the right eye, the corneoscleral border was obscured, partly because the limbus was irregularly broadened and partly because the cornea was opacified in this region. The cornea was smaller than normal, was flattened, and bore flecks at points which were attached to filaments from the iris. The angle of the anterior chamber was undeveloped, and the chamber shallow. The iris arose directly from the opaque lim-

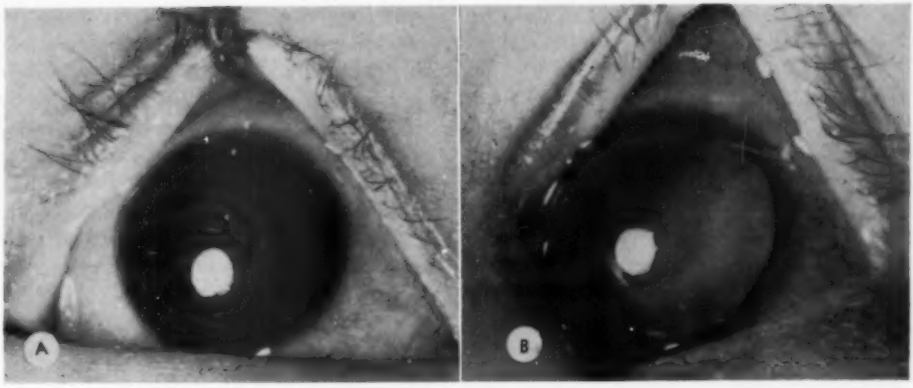

Fig. 2 (Cole and Cole). (A) Right eye. (B) Left eye before enucleation.

bal zone, was poor in stroma, with irregular pigmentation. Toward the periphery the stroma formed thickened strands with irregular pockets between them. Behind the lens, which as far as it could be seen was transparent, lay a structureless mass, gray-white in color, and without vessels. Since the mass could not be transilluminated, no further examination was possible. This lesion was of the type called by the Americans retrolental fibroplasia.

4. Scott, Friedmann, Chitters, and Pepler,[6] 1955: At eight months it was noted that the child could no longer recognize her parents. Nystagmus was present in both eyes. The right pupil was dilated and fixed, the vitreous hazy, and the disc pale, but no other abnormalities were present at this or later examinations. The left eye was shrinking, and fundus examination suggested metastatic endophthalmitis. At the age of one year, a gray, smooth mass was clearly visible in the posterior part of the eye, with a red reflex around the edge. By the age of 18 months the mass had increased in size until it lay directly behind the lens. Cortisone was tried, but was not beneficial. Because of the steady growth of the mass, the eye was enucleated.

The pathologist reported: "On section the most striking feature was a completely detached, funnel-shaped, folded, and thickened retina which showed patchy infiltration by lymphocytes. On the medial side only, the detachment involved the pars plana and the ciliary body. Lying between the lens and the detached retina was a thick, fibrous, cellular membrane which was attached to the pars plana retinae on each side. The space behind the retina contained albuminous fluid in which numerous epithelial cells (ghost cells) and fatty acid crystals could be seen. The

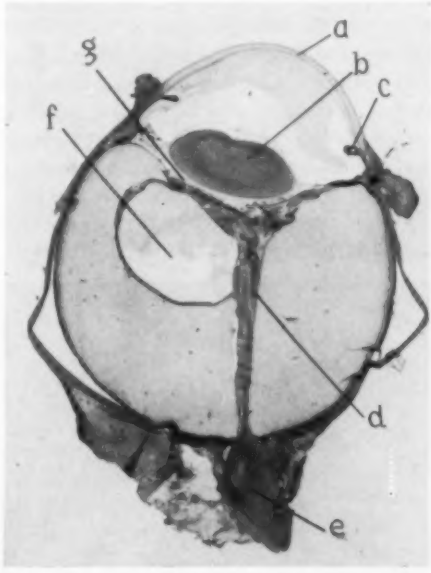

Fig. 3 (Cole and Cole). Sagittal section through enucleated eye. (a) Megalocornea. (b) Ectopia lentis. (c) Cyst of the iris. (d) Detached retina. (e) Optic nerve. (f) Cyst of retina. (g) Retrolental fibroplasia or ablatio falciformis.

cornea, the anterior chamber, and the lens, though slightly distorted by sectioning, appeared healthy. The iris showed a large posterior synechia, but no pathologic changes could be observed in the stroma. There was some edema and slight infiltration of lymphocytes and an occasional polymorphonuclear lymphocyte toward the cut end of the optic nerve."

5. Findlay,[11] 1956: The left eye was small and did not respond to light. Fundus examination showed that the disc was white and normal in size. The retinal and choroidal bed was however grossly dilated and suggested streaks of hemorrhage radiating from the disc; punctate hemorrhages were visible at other points. Elevated grayish areas were seen at the periphery at the 3-, 6-, and 9-oclock positions, that at the 6-o'clock position extending eight mm. toward the equator. The ciliary body was not seen, the vitreous was clear, and the anterior chamber rather shallow. Penicillin had no effect on either the skin or the eye lesions, but cortisone caused a temporary recession of the skin lesion. Four months later the mass at the 3-o'clock position had reached the nasal side of the lens and had fused with that at the 6-o'clock position. The changes were considered typical of retrolental fibroplasia. The mother, however, had a similar eye condition and a skin defect which suggested a sex-linked genetic factor. This and the asymmetry in the two eyes and the term birth suggested that ablatio falciformis might be the more correct diagnosis.

6. Jensen,[12] 1956: The right eye was normal. The left showed convergence and severe amblyopia. A large, oblique, whitish membrane with vascularized processes extended into the vitreous to the retina, the more voluminous portion hiding the optic nerve. The mebrane suggested a proliferation of connective tissue. The part of the retina adjacent to the mass was slightly depigmented, and more peripherally there were elongated but poorly defined accumulations of dense pigmentation. At the extreme periphery were slight diffuse pigment changes and two small areas of retinal hemorrhage. A layer of blood was seen at the lower posterior surface of the lens. Both mother and child reacted positively to toxoplasmosis tests. The diagnosis was an ocular complication of incontinentia pigmenti of the type variously called pseudoglioma, retrolental fibroplasia, metastatic ophthalmia, or absence of vitreous.

7. Lieb and Guerry,[7] 1957: The external eyes were both normal. Examination of the fundus of the right eye showed a normally colored disc with sharp edges, with a small pigment sickle at the temporal side. The superior and inferior temporal veins were strikingly engorged, the vessels above resembling pannus vascularization, and one branch below extremely tortuous. The retina between these vessels was markedly edematous, especially in the areas of intensive vascularization. Extensive pigment changes were pres-

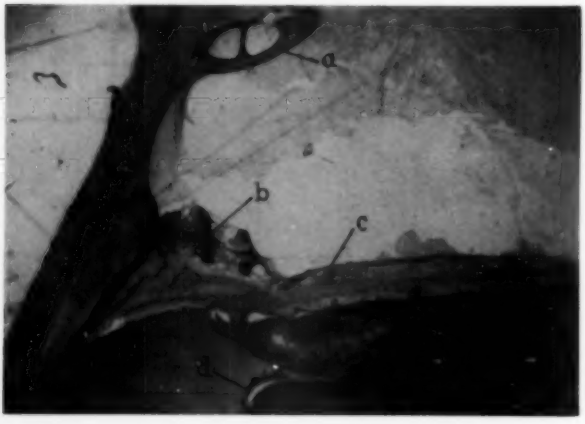

Fig. 4 (Cole and Cole). Photomicrograph showing the site of the membrane. (a) Cyst of the iris. (b) Ciliary body. (c) Retrolental membrane. (d) Detached retina.

ent in the macular area, and the fovea was covered by a kidney-shaped whitish area and by exudate. Above the macular area in the vascularized region lay a large elevated whitish form; while the lower border of this exudative area was sharp, the upper shaded off into general edema. In the surrounding area there were several star-shaped masses of whitish exudate. In the left eye, extensive edema and tortuous vessels were present, but no other abnormalities. The diagnosis was exudative chorioretinitis.

11. Cole, 1958: The child could distinguish light, but had no following movements. Examination of the right eye showed a shallow anterior chamber; the iris was bound to a cataractous, yellow lens which transilluminated well from all angles, but prevented examination of the posterior chamber. The left eye had an enlarged and somewhat hazy cornea and a deep anterior chamber. Elongated ciliary processes were inserted into the retrolental space. Central to the ciliary area was a circular area of hemorrhage. Behind the lens was a yellowish membrane with no vessels on the surface. Transillumination revealed small areas at the center and below, which appeared dark when lighted from any angle. Tension in the right eye was 20 mm. Hg and in the left 27 mm. Hg (Schiøtz). Because the growth in the left eye strongly suggested glioma, the eye was enucleated. Hoping that some vision might be present in the right eye, an iridectomy and lens needling were performed, but a mass was found in this eye also.

The pathologist reported a bulging cornea, normal except for decrease in the thickness of the substantia propria. The limbus was edematous and contained cellular exudate. The greater part of the iris angle was obliterated by adhesion of the iris over most of the limbus, but the marginal sinus was clearly identified and included cysts in some areas. The iris was atrophied and distorted, and the pigment lay several millimeters over the posterior surface. On one side a newly formed membrane extended from the iris to the cornea. The ciliary body and ciliary processes were deformed by traction on the posterior surface of the lens. In addition, a membrane had formed on the ciliary body due to chronic cellular exudate and fibrin deposition.

Fig. 5 (Cole and Cole). Higher-power view of the membrane above and the folded retina below.

Except for some thinning, the choroid and sclera were not altered except in the area near the nerve where there was considerable inflammatory reaction which extended through the cribriform plate into the vaginal sheath. The pigment epithelium showed some degeneration at the ora serata and near the optic nerve.

The retina was completely detached and incorporated into a membrane which extended across the retrolental space and also extended posteriorly to the optic nerve, which it tended to pull forward. The membrane was made up of proliferating fibrous tissue containing new vessels and evidence of hemorrhage in the posterior extension. The normal cellular structure of the retina was hidden in this massive fibrous detachment and

by a cyst which separated its layers on one side.

The cyst measured five mm. anteroposteriorly and eight mm. transversely. Both the cyst and the space back of the retina were filled with clear, pink-staining fluid containing some debris. The diagnosis of the basic anomaly was retrolental fibroplasia or ablatio falciformis.

To these eight cases should probably be added two more which are only meagerly described: (1) Kitamura's Case 19 in which a blind left eye was enucleated with a postoperative diagnosis of retrolental fibroplasia, and (2) Kitamura's Case 20 in which a condition in the left eye was diagnosed as metastatic ophthalmia and the eye later became blind;[13] and possibly a third, (3) Seidelmayer and Moncorp's[14] case in which atrophy of the optic nerve was considered due to glioma. This diagnosis cannot be verified and remains questionable, but is of interest in relation to other diagnoses. Judging from the family histories of some patients, the incidence of this lesion is probably higher than is indicated by this group, but data on other cases are not sufficient to warrant their inclusion in this survey.

Discussion

The most conspicuous fact in this series is that in one-half of the eight cases, and one of the additional three, an eye was enucleated, apparently because the ophthalmologist could not certainly rule out glioma or other neoplasm on clinical grounds. Moreover, in one other case glioma was postulated. The same problem has also been noted in other cases with similar congenital eye dysplasias.[7,15-17] As far as can be ascertained, the diagnosis of neoplasm has not been verified in any of these cases, whether or not they were associated with the incontinentia pigmenti syndrome. The problem seems to be particularly acute when, as in most of our cases, the mass was unilateral and the other eye healthy or relatively little affected.

Second, while the lesions seem related, descriptions of details vary considerably, but perhaps not more than is to be expected among congenital lesions. Moreover, since this particular lesion is very rare, observations and interpretations of different investigators may well vary more than if they were reporting better known syndromes. For the same reason, probably diagnoses often constitute descriptions in terms of better understood comparable lesions rather than an intention of final diagnosis. We therefore need a somewhat firmer ground for diagnosing these masses, and especially for distinguishing them from neoplasms.

The less severe of these masses suggest the lesions described by Mann[15] as congenital retinal fold. Mann also noted that these cases tended to overlap with other somewhat similar ones with retinal detachment and microphthalmos. Weve[16] described the same syndrome somewhat more fully, and included more severe lesions under the name of ablatio falciformis. Weve found a genetic factor in many of his cases, and noted that lesions were bilateral in one half of the total 22. His histologic studies suggest a strong resemblance to the histologic reports of eyes enucleated for mass associated with incontinentia pigmenti syndrome. Weve pointed out the resemblance of some of these cases to retrolental fibroplasia.

Reese[17] on the basis of careful analysis of pathologic specimens and study of the literature found that congenital eye defects described under a wide variety of terms could actually be classed as of two kinds: (1) persistence and hyperplasia of the primary vitreous, and (2) retrolental fibroplasia. In the first type, the retina contains all its elements and is not detached, at least not in the essential visual areas. In the second, the retina is not fully differentiated, an abnormality exists in the subretinal layers with evidence of at least a mild preceding choroiditis, a persistence of primary vitreous is present at the base of the lesion, and the retina—except in early stages—is detached to a greater or lesser degree.

For more complete descriptions of the clinical and pathologic differences the reader is referred to the original paper. As far as can be ascertained from the report, advanced

forms of the two kinds of lesions may be somewhat difficult to distinguish both from each other and from neoplasms. Reese believes that retrolental fibroplasia includes syndromes described as congenital retinal fold, ablatio falciformis congenita, metastatic retinitis, intrauterine endophthalmitis, opaque membrane behind the lens, shrunken fibrous tissue cataract, congenital connective tissue formation in the vitreous, and fibrous tissue cataract.

A mass in the posterior chamber associated with incontinentia pigmenti differs from typical retrolental fibroplasia, however, in several ways. It is a congenital lesion, occurs usually in term babies, and is most frequently unilateral. Although the problem of congenital malformation is as yet only poorly understood, the multiplicity of possible causes for dysplasia is well known, and includes such diverse factors as infection, heredity, accident during the period of gestation, and vitamin or other nutritional deficiencies. Undoubtedly the variety of possible prenatal insults, as well as differences in the severity and duration of the insult, accounts for the fact that the masses occurring in the posterior chamber of the eye in cases of incontinentia pigmenti are clinically more variable than those of retrolental fibroplasia and are also more likely to be associated with other congenital eye defects.

In the incontinentia pigmenti syndrome there is some evidence that inflammation may play an important role at least in some instances of mass in the posterior chamber. Lieb and Guerry's case, with a definite raised area in the retina, was diagnosed as chorioretinitis. One of Weve's patients had been similarly diagnosed previously, and in the histories of some of his other cases, relatives had "pseudoglioma," chorioretinitis, or ablatio falciformis. The fact that the characteristic skin defect is usually preceded by an inflammatory eruption is consistent with this view. Similar observations have been made by other authors.[12]

With this information it should not be difficult to differentiate mass in the posterior chamber associated with incontinentia pigmenti syndrome from neoplasm. The more severe lesions are very likely to be associated with other defects, or at least with disturbed metabolism.[18] In some few instances the presence of eye or other congenital anomalies in the child's family may provide a clue.[19,20]

Conclusion

1. The incontinentia pigmenti syndrome usually includes associated congential defects beside those of the skin.

2. In about one third of cases the dysplasia is manifested in one eye, sometimes in both eyes, but rarely to the same degree in both eyes.

3. At least one third of the ocular defects involve mass in the posterior chamber. These masses seem difficult to differentiate from retrolental fibroplasia. They are, however, congenital, are somewhat more variable, and more likely to be associated with other eye defects.

4. If the incontinentia pigmenti skin lesion occurs, it is advisable to search carefully for other anomalies.

5. The more severe cases of mass in the posterior chamber can usually be differentiated from neoplasm by the presence of other congenital anomalies or defects in metabolism, and occasionally by the presence of similar defects in the family, suggesting a genetic factor. As far as is known, provisional diagnosis of neoplasm has not been established in any of these cases.

780 Park Avenue (21).

Acknowledgment

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DACRYOCYSTORHINOSTOMY*

A REVIEW OF 106 OPERATIONS

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Dacryocystorhinostomy is a procedure done infrequently and with such a variety of techniques on the average ophthalmologic service that most ophthalmologists seem unaware of the incidence of success and failure or the incidence of complications that may occur with any given technique.

A review of the American literature reveals few recent articles on the results of all types of surgery for chronic dacryocystitis. Prior to the present century, surgery of the lacrimal apparatus was limited to operations for tumors of the lacrimal gland, and few ophthalmologists attempted surgery on the sac.[1] Epiphora was treated by probings and suppurative dacryocystitis by irrigation with antiseptic solutions and occasionally cauterization of the sac. At the end of the past century and for a period of several years extirpation of the sac was done and was generally considered the best operation for chronic dacryocystitis.

In 1904, Toti described his procedure which consisted of draining the contents of the lacrimal sac directly into the nasal fossa through an opening in the nasal bone with excision of the nasal wall of the sac and the lateral nasal mucosa.[2] This operation was used infrequently because of the low inci-

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dence of successful results.[3] Improvements of the technique of Toti were described by Dupuy-Dutemps and Bourguet.[4] These consisted of suturing the mucosa of the sac directly to the nasal mucous membrane without excision of either part of the sac or nasal mucosa.

Intranasal dacryocystorhinostomy was first described by West in 1910 and since has been used by rhinologists but has never been widely accepted by ophthalmologists.[5] Mosher has described a modification of the Toti technique which combines the external and intranasal techniques, and this, too, has been used by both ophthalmologists and rhinologists.[6]

For the past 25 years most operations for chronic dacryocystitis have been performed by ophthalmologists and, for the most part, these have consisted of some modification of the Dupuy-Dutemps technique of external dacryocystorhinostomy.[7-9]

We have recently had the opportunity to review our results with dacryocystorhinostomy, and it is the purpose of this paper to present the results of 106 operations.

Clinical Material

The clinical material for this study was derived from the case histories of all patients admitted to McPherson Hospital and the University of North Carolina Memorial Hospital with the diagnosis of chronic dacryocystitis. Those case histories of patients with dacryocystitis on whom dacryocystorhinostomy had been performed and in which adequate follow-up was available for the determination of success or failure were used for study. There were 99 such histories available; 106 operations were performed on these patients, 99 operations were done as primary procedures and seven operations as secondary procedures.

The age distribution of patients at the time of operation is shown in Figure 1. Sixteen operations were done in persons less than 30 years of age; 32 operations were done on persons 30 to 50 years of age; 49 operations were done on persons 50 to 70 years of age; nine operations were done on persons over 70 years of age.

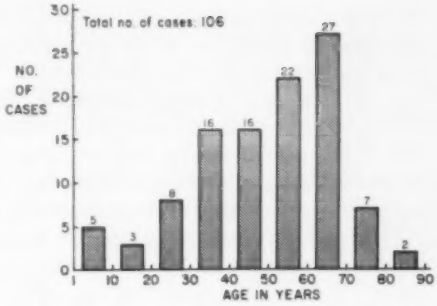

Fig. 1 (McPherson and Egleston). Age distribution at time of operation.

Ninety-three of these operations were done on white and 13 on Negro patients; 83 of these operations were done on females and 23 on males.

Etiology

The probable causes of dacryocystitis are shown in Table 1. In 87 cases no cause could be determined except for chronic infection of the sac. In seven cases the disorder was considered to be congenital in origin. Four cases were thought to be due to trauma; five cases to aberrant ethmoid cells encroaching on the lacrimal fossa; and one case to a mucocele of the antrum. In one case a bony tumor in the nose encroached on the lacrimal fossa. One case was associated with a saddle-nose deformity of congenital syphilis.

Technique

The technique used in the present series was essentially that of Dupuy-Dutemps. Anesthesia is obtained by infiltration of the area over the sac with two-percent procaine with adrenalin; blocking the supraorbital, supra-

TABLE 1

Probable Etiology of Dacryocystitis

Etiology	
Unknown	87
Congenital	7
Trauma	4
Ethmoid cells	5
Mucocele of antrum	1
Bony tumor of nose	1
Syphilis	1

trochlear, infratrochlear, anterior ethmoidal, and infraorbital nerves. A pledget saturated with four-percent cocaine and adrenalin is packed in the middle nasal meatus. The skin is incised in a crescentic manner nasal to the fossa. The medial canthal ligament may be incised at its insertion into the nasal bone if necessary for exposure. The internal wall of the sac and periosteum are reflected exposing the lacrimal fossa. The anterior lacrimal crest is perforated by means of a dental burr or Stryker saw with the Iliff trephine, and the opening is enlarged to the posterior lacrimal crest with Kerrison forceps and rongeurs.[10] An ovoid opening is made which is approximately 1.5 by 1.5 cm. A probe is passed through the lower punctum into the sac. With this as a guide, a vertical incision is made into the sac, and short horizontal incisions are made at the extremities of the first incision. Similar incisions are then made in the opposite nasal mucosa. The posterior leaf of the sac is sutured to the posterior leaf of the nasal mucosa with two catgut sutures. A rubber catheter is then passed through the sac and out the corresponding naris. This is secured to the anterior leaf of the sac and to the anterior leaf of the nasal mucosa with two additional catgut sutures (fig. 2). The subcutaneous tissues are approximated with interrupted catgut sutures, and the skin incision is closed with subcuticular Dermalon. Irrigation of the lower punctum is performed, and an antibiotic ointment and pressure dressing applied.

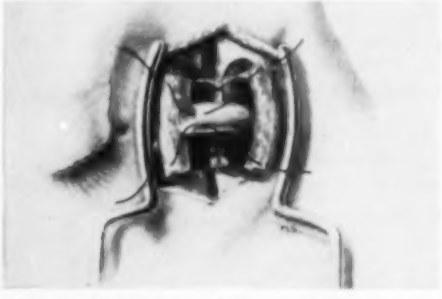

Fig. 2 (McPherson and Egleston). Technique of suturing rubber catheter to anterior mucous membrane flaps.

Results and Complications

The results of these operations are shown in Table 2. Operation was considered successful if, after good wound healing, the patient was symptomatically relieved and irrigation into the nose was easily performed. Of the 99 primary operations performed, 84 were successful by these criteria; 15 were considered failures. Of the 15 failures, seven persons consented to secondary operation; five of these were successful and two were failures. Eight patients refused secondary operation. The average time of failure was found to be six weeks postoperatively. The earliest failure occurred six days after operation and the latest 12 weeks. In one case acute dacryocystitis recurred six years after a successful primary operation.

The operative complications encountered were few. Bleeding was occasionally brisk from subcutaneous tissues and oozing from bone bleeders was almost routinely encountered. On two occasions the angular artery was severed. It was found that this could be avoided by placing the incision one cm. nasal to the lacrimal fossa. Occasionally the operator noted difficulty in determining the exact lumen of the lacrimal sac when it was opened. This was particularly noted when infection had been present for some period of time and the wall of the sac was greatly thickened.

The immediate postoperative complications consisted of one instance of severe postoperative epistaxis occurring on the third postoperative day and one of bleeding into the subcutaneous tissues on the second postoperative day. There was one instance of stitch abscess and one of postoperative sac abscess.

TABLE 2

Results of primary and secondary operations

Results	Primary Operation	Secondary Operation*	Total
Success	84	5	89
Failure	15	2	17
Total	99	7	106

* Eight patients refused secondary operation.

The late postoperative complications were those associated with closure of the rhinostomy with recurrence of epiphora. An analysis of the 15 failures reveals no common predisposing factor. There was no correlation of failure with duration of disease, age of patient, sex, or race.

Of the seven patients who were reoperated, three were found to have dense scar tissue present in the rhinostomy. One patient was found to have a bone spur projecting into the rhinostomy, two patients were found to have septate sacs, and one patient developed a postoperative adhesion between the middle turbinate and the lateral nasal wall occluding the new opening.

Discussion

It is difficult to account for the failures which occur following dacryocystorhinostomy. The lack of correlation of failure with age, sex, or race distribution of patients indicates that failure is probably due to technical error at the time of operation. The exact nature of this error is not clear. The low incidence of postoperative hemorrhage or infection indicates that this error may be mechanical and may occur in performing the rhinostomy or in suturing the mucous membrane flaps. In 43 of the 106 operations, the Iliff trephine was used and in 63 a dental drill. Although the trephine gives a cleaner rhinostomy, there was no difference in the incidence of success in cases in which the trephine was used as compared with those in which the drill was used. The mucous membrane flaps were sutured in essentially the same manner in all cases.

The difficulty in determining the exact lumen of the lacrimal sac, which was occasionally noted in the operative record, may be a factor in the failures which occurred. The finding of two instances of divided sacs at reoperation indicates that in at least these two cases the operator may have sutured nasal mucosa to sac wall and not completely to sac lumen.

The inability to demonstrate a common factor to account for failure in these 15 cases indicates that dacryocystorhinostomy requires meticulous attention to technical detail by the surgeon if it is to be routinely successful.

These details may include careful placement of the incision to avoid the angular vessels, atraumatic handling of soft tissues, a clean rhinostomy, careful suturing of mucous membrane flaps, and careful dissection to expose the true lumen of the sac.

Summary

1. The results of 106 dacryocystorhinostomies are presented.

2. Primary operation was successful in 84 of 99 operations.

3. Secondary operation was successful in five of seven operations.

4. Fifteen unsuccessful operations are analyzed and no common factor was found to account for failure.

5. If routine success of dacryocystorhinostomy is to be attained, meticulous attention to technical detail by the operating surgeon is necessary.

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THE ASSOCIATION OF KERATOCONUS WITH ATOPIC DERMATITIS*

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The relation of atopic dermatitis (neurodermatitis disseminata) to various manifestations of ocular disease has been well documented in the ophthalmologic literature. Cataract formation,[1-14] retinal detachment,[6, 15, 16] and keratoconjunctivitis[17] have been noted most often. Keratoconus is apparently uncommon but has also been reported, first (1937) by Hilgartner, Hilgartner, and Gilbert,[18] who found it in a patient with a history of allergic symptoms suggesting atopic dermatitis and then (1942) by Bereston and Baer[19] who found it in two atopic individuals. The latter investigators felt that in both their cases the two conditions might very well be intimately associated but that the possibility of coincidence could not be excluded. Evidence that the connection may be more than coincidental is found in an article by Brunsting, Reed, and Bair[5] who noted six cases of keratoconus in a series of 1,158 atopic patients; four of the six were females and all were between the ages of 12 and 23 years; the keratoconus was bilateral in all six, and three of the six had lens opacities.

The purpose of the present report is to describe the six cases of bilateral keratoconus that have been seen by us in the Eye Clinic of the University of California Medical Center during the past three years. All six were in atopic patients. Four of the six were females, all were white, and all were between the ages of 20 and 32. One also had a unilateral cataract, three had an associated keratoconjunctivitis, and one had a unilateral retinal detachment.

CASE REPORTS

CASE 1

L. B., a 20-year-old housewife, had a history of eczema as an infant, asthma at four years of age, and recurrent attacks of dermatitis since puberty. A diagnosis of atopic dermatitis was made by the referring physician.

This patient first noted poor vision at the time of her first pregnancy, 17 months prior to her examination at the eye clinic. Her vision had improved with corrective lenses until seven months prior to examination when they no longer helped. Contact lenses were then prescribed. These improved her vision somewhat but she was unable to wear them because of recurrent attacks of itching of the eyes associated with stringy discharge. Three weeks prior to examination she awoke with a stabbing pain in her right eye.

Examination. Visual acuity was correctible in the right eye to counting fingers at two feet, and in the left eye to 20/100. The skin of the lids was thickened. The conjunctiva was moderately thickened. There was an associated shrinkage of the lower cul-de-sac and a papillary hypertrophy of the upper and lower tarsal conjunctiva. The right cornea showed marked keratoconus with central thinning, bullous keratopathy, and a healing central epithelial erosion. The left cornea was conical and displayed early striate keratopathy. Keratometer readings showed a marked irregularity of the mires. (We are indebted to Dr. Phillips Thygeson for the opportunity to examine this patient and to include the findings in this report.)

CASE 2

D. C., a 28-year-old laborer, had a history of allergy and skin difficulties as long as he could remember.

This patient had recently been treated at the dermatology clinic for exacerbations of his atopic dermatitis, and at the Langley Porter Psychiatric Clinic for an associated psychoneurosis. Four years previously he had been seen at the eye clinic. At that time he stated that he had lost the vision in his right eye at the age of three years after a spontaneous retinal detachment, and that he had been seen at that time by an ophthalmologist who advised against surgery.

Examination (June, 1954). Vision in the right eye, no light perception; vision in the left eye, 20/70 without correction. The lids were thickened, the conjunctivas normal. There was bilateral keratoconus, more marked in the right eye than in the left. The right cornea had a small central leukoma and was quite thin centrally. The left cornea was conical but there was no opacification. The vision of the left eye could be improved with a spectacle lens to 20/40 and with a contact lens to 20/15. A dense cataract of the right eye prevented a view of the right fundus. The left fundus was normal.

The patient was not seen again until November 10, 1957, when he presented himself at the emer-

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gency room stating that he had awakened with a painful left eye.

Examination (November, 1957). The right cornea displayed an epithelial erosion, bullous keratopathy, marked thinning of the cornea, and a beginning descemetocele. The patient was hospitalized. After local therapy with hypertonic solutions and mydriatics, plus systemic steroid therapy for a concurrent exacerbation of his dermatitis, a full-thickness 8.0 mm. keratoplasty was performed without incident.

CASE 3

C. C., a 20-year-old housewife, had a history of asthma as a child and skin difficulties since the age of 12. This patient had been treated at the dermatology clinic of this hospital for a severe exacerbation of her dermatitis which occurred at the time of her first pregnancy in January, 1955. The skin difficulty was diagnosed as atopic dermatitis.

The patient first noted a decrease in visual acuity at the time of her pregnancy in 1955. Glasses prescribed were unsatisfactory. The patient was first examined at the University Eye Clinic in the fall of 1956.

Examination. Vision in the right eye, 20/200; vision in the left eye, 20/70; correctible to R.E., 20/70; L.E., 20/30, with spectacle lenses, and to R.E., 20/50; L.E., 20/20, with contact lenses. The skin of the lids was thickened, erythematous, and scaly. The conjunctiva was thickened and there was a papillary hypertrophy of the upper and lower tarsal conjunctiva. A bilateral keratoconus was noted, with marked distortion of the mires of the keratometer in the right eye, moderate distortion in the left. The right cornea showed central thinning, a conical shape, and small folds of Descemet's membrane centrally. The left cornea was free of opacities.

CASE 4

I. M., a 26-year-old accountant, was first seen in the eye clinic in July, 1950, with a history of decreasing vision in both eyes for four years. A diagnosis of keratoconus was made by the referring physician. The patient also stated that he was being treated by his dermatologist for atopic dermatitis, and that he had had asthma as a child.

Examination. The skin of the lids was thickened and redundant, giving the patient the appearance of having "bags" under his eyes. The conjunctiva was normal in appearance. Both corneas were conical and thinned centrally. There was a linear defect in Descemet's membrane centrally in the left eye. The vision in each eye was 20/30 when corrected with spectacle lenses. This was improved to 20/20, O.U., with contact lenses. Keratometer readings taken at yearly intervals revealed a slow progression of the degree of corneal astigmatism. In 1950 there were 3.0 diopters of astigmatism in the right eye and 3.5 diopters in the left; in 1957 there were 4.0 diopters in the right eye and 4.5 in the left.

CASE 5

J. T., a 24-year-old housewife, was first seen in July, 1956, with a history of keratoplasty of the left eye performed at another hospital in May, 1956. The patient moved to this city because of her "allergies" and requested care at the University Eye Clinic. Bilateral keratoconus had first been noted at 16 years of age. Over the years she had been treated by her family physician for hay fever, occasional bouts of asthma, and recurrent eczematoid dermatitis.

Examination. There was a thickening of the skin of the lids and a bilateral papillary hypertrophy of the upper and lower tarsal conjunctiva. An eosinophilia was noted in scrapings from the conjunctiva. The right cornea was thinned centrally and showed striae of Descemet's membrane and epithelial bullae. The left cornea presented a clear 7.5-mm. full-thickness graft which appeared normal in all respects. The vision in the right eye was limited to finger counting at four feet; the vision in the left eye was correctible to 20/30.

On November 14, 1957, a 7.5-mm. full-thickness keratoplasty was performed on the right eye. The postoperative course has been normal to date.

CASE 6

E. V., a 32-year-old secretary, had a history of recurrent attacks of eczema and asthma for many years, made worse by emotional tension. This patient had been treated for the previous five years at the dermatology clinic for atopic dermatitis. Her vision had been poor since the age of 18 years.

Examination. The skin of the lids was dry and thickened. The conjunctivas were normal. Both corneas were conical, the left markedly so. Vision was reduced to 20/80 in the right eye, and to 20/200 in the left eye, with and without correction. There was an early plaquelike opacity in the anterior cortex of the right crystalline lens. Refraction: R.E., +1.75D. sph. ⊃ +1.5D. cyl. ax. 140°, 20/40; L.E., −3.5D. sph. ⊃ +11.5D. cyl. ax. 30°, 20/40.

The patient was examined at intervals of six months. She noted progressive loss of vision in the right eye due to maturation of the cataract. In January, 1953, the vision in the right eye was reduced to light perception. An uneventful intracapsular cataract extraction was performed on the right eye, and the patient was able thereafter to read 20/20 with this eye, wearing +11.5D. sph. ⊃ −2.5D. cyl. ax. 135°. She has been seen periodically since the operation and there has been very little change in her refractive error. A refraction in October, 1957, revealed a 4.0D. cyl. in the right eye and a 12.0D. cyl. in the left.

DISCUSSION

The association of lens opacification and retinal detachment with atopic dermatitis has been considered as suggestive evidence that only structures of ectodermal origin are involved in the atopic process[6-11] but, if keratoconus and the keratoconjunctivitis described by Hogan[17] are also part of the atopy, this theory would appear to be untenable.

Many of the manifestations of atopic dermatitis cannot, in fact, be interpreted as reflections of a strictly ectodermal defect. That there is also mesodermal involvement is indicated by the vascular abnormalities noted in atopic individuals by Weber, Roth, and Kierland,[20] by the phenomenon of white dermographism, and by the fact that atopic patients usually exhibit a high degree of allergic sensitivity. Actually the dermis is the "shock organ" in the skin, and the dermis, like the major portion of the cornea, is of mesodermal origin.

It is noteworthy that three of our patients displayed papillary hypertrophy of the conjunctiva of both the upper and lower lids. This picture, in association with a stringy discharge and conjunctival eosinophilia, could suggest vernal conjunctivitis. As Thygeson[21] has pointed out, however, papillary hypertrophy of the lower tarsal conjunctiva is in fact so unusual in vernal conjunctivitis as to constitute a useful differentiating point between the latter condition and atopic involvement of the conjunctiva.

Summary and Conclusions

1. In a three-year period, the six cases of keratoconus seen by us at the University of California Eye Clinic were in patients with atopic dermatitis. Four of the six were females, one had a unilateral cataract, three had an associated keratoconjunctivitis, and one had a unilateral retinal detachment.

2. If keratoconus can be associated etiologically with atopic dermatitis, as is strongly suggested by this group of cases, the atopic process cannot be regarded as limited to structures of ectodermal origin. The same conclusion can be drawn from the etiologic association of keratoconjunctivitis with atopic dermatitis (Hogan[17]).

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SECONDARY GLAUCOMA FOLLOWING OCCLUSION OF THE CENTRAL RETINAL ARTERY*

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Secondary glaucoma following occlusion of the central retinal artery can no longer be considered a rare occurrence. The fact that this clinical entity has found so little attention in the literature is hard to understand. We believe that many of these cases must have been overlooked by eye pathologists and misdiagnosed as "hemorrhagic" secondary glaucoma following occlusion of the central retinal vein. This latter entity is much more common and exhibits in its late stages very similar histologic findings—except for the fact that not the artery but the vein shows the occlusion.

Only six clinically and pathologically well-documented cases of secondary glaucoma following occlusion of the central retinal artery were found in the literature. These cases were reported by Opin,[1] Bussola,[2] Benton,[3] Wolter and Lubeck,[4] and Wolter and Liddicoat.[5] We know that both Teng in New York and Zimmerman at the Armed Forces Institute of Pathology in Washington have also seen cases of this entity and are going to publish their studies of these cases (personal communication).

The present paper represents a very complete clinical and histologic demonstration of a case of this entity. The observation of the arterial occlusion and the development of the secondary glaucoma as well as the histologic study were done continuously by the same examiners. The history and the findings of this case leave no doubt that the entity of secondary glaucoma following occlusion of the central retinal artery really exists.

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Case history

This 59-year-old man was admitted to Wayne County General Hospital on May 25, 1955, for convalescence and custodial care after a cerebral vascular accident caused by essential hypertension. Blood pressure on admission was 250/150 mm. Hg. On June 8th, the patient was seen in the ophthalmology department for evaluation of the fundi. His visual acuity was found to be: O.D., 20/25; O.S., 20/50. The intraocular pressure was normal, O.U. Both fundi exhibited a grade II hypertensive (angiospastic) retinopathy (Keith-Wagner) without hemorrhages or exudates. His visual fields were normal. The medical treatment consisted of Serpasil and large doses of Ansolysin which brought his blood pressure very gradually over a two-year period down to 126/70 mm. Hg.

The patient was again seen by us on January 28, 1957, at which time his fundi were again described as a grade II hypertensive retinopathy without hemorrhages or exudates. The patient continued on his antihypertensive regime and was not seen until March 13, 1958. At this time he came because of a marked loss of vision in his right eye for one week.

His visual acuity was: O.D., light perception; O.S., 20/50. His right pupil showed poor reaction to light. The intraocular pressure, O.U., was normal. The fundus of the right eye showed the disc to be slightly pale and of poor vascularity. The arterioles appeared very small and white for two to three disc diameters and showed no apparent blood flow. The macula appeared quite red in contrast to the surrounding retina. There were no hemorrhages or exudates. The left eye again presented a grade II hypertensive retinopathy and some arteriosclerotic vascular changes. The diagnosis of central retinal artery occlusion, O.D., was made.

On April 20, 1958, the patient was admitted for an attack of acute appendicitis but at surgery was found to have regional ileitis. The postoperative course was uneventful. On May 13, 1958, he was again seen because of an inflamed, "sore" right eye for two days. This time he was found to be completely blind in his right eye; visual acuity, O.S., was still 20/50. His right eye had redness and swelling of the lids and marked conjunctival and pericorneal injection. The cornea was steamy and showed some small central bullas. The pupil was moderately dilated and the iris had early rubeosis iridis. The fundus could not be visualized. The intraocular pressure in his right eye was 60 mm. Hg. The left eye was unchanged from the previous examinations and had a normal intraocular

pressure, and a deep anterior chamber.

The diagnosis of secondary glaucoma following occlusion of the central retinal artery was made and the patient was started on large doses of Diamox and pilocarpine. This resulted in little or no change in the intraocular pressure. The corneal bullas and the rubeosis iridis increased, as did the patient's discomfort. On May 22, 1958, the right eye was enucleated and immediately fixed in bromformalin (Cajal solution). The postoperative course was uneventful.

Histologic examination

Method

Half of the globe was imbedded in paraffin, cut in serial sections, and stained with routine methods of eye pathology. The whole optic nerve was imbedded separately in paraffin and cut in serial cross sections which were stained with hematoxylin and eosin. Frozen sections were made of the retina, choroid, and ciliary body of the other half of the globe. These latter sections were stained with the silver carbonate methods of Rio Hortega as described in detail by Scharenberg and Zeman.[6]

All illustrations presented in this paper are unretouched photomicrographs.

Results

The eye was of normal size and shape. The anterior chamber was shallow. The lens was clear and in its normal position. No retinal hemorrhages were seen macroscopically. The vitreous was liquefied.

The microscopic examination revealed the cornea to be normal except for a few intraepithelial bullas in its center. There was complete closure of the anterior chamber angle by extensive peripheral anterior synechias and proliferated fibrovascular tissue. The trabecular system showed advanced degeneration of its endothelium, swelling of the trabecular meshwork and deposition of pigment. The iris exhibited a continuous layer of newly formed, thin-walled blood vessels on its anterior surface (fig. 1). The structure of the ciliary body appeared normal after routine staining. The silver stains, however, revealed most of the nerve fibers of the ciliary plexus to exhibit very severe degeneration (fig. 2). These nerve fibers were swollen and interrupted and many of them showed a peculiar shape like a string of pearls. The choroid was normal and it must be emphasized that the choroidal arteries and veins were very well preserved and showed only very little sclerosis. The pigment epithelium was normal.

Extensive degeneration was found all through the retina (fig. 3). This degeneration mainly involved the inner retinal layers. The ganglion cells, nerve fibers, and astroglia of these inner layers were virtually missing. Only the brushlike inner processes of the radial fibers of Mueller and the blood vessels were left of the inner retinal layers (fig. 4). The lumen of all the arteries were virtually obliterated and the walls very much thickened. The veins were open but also showed thickened walls and small lumen. No hemorrhages or exudates were found in the retina. The outer layers with the rods and cones were surprisingly well preserved.

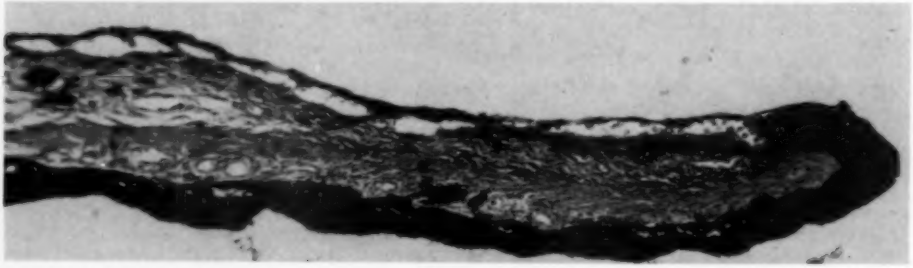

Fig. 1 (Wolter and Phillips). The iris shows a continuous layer of new-formed, thin-walled blood vessels on its anterior surface (rubeosis iridis). (Hematoxylin-eosin, photomicrograph.)

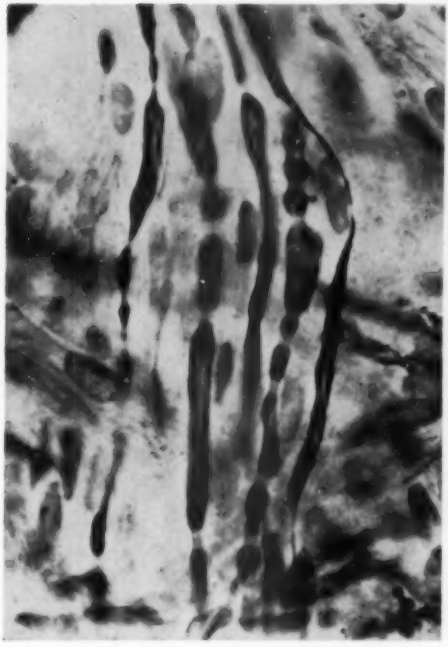

Fig. 2 (Wolter and Phillips). Bundle of nerve fibers of the ciliary body which show advanced degeneration. (Hortega method, photomicrograph.)

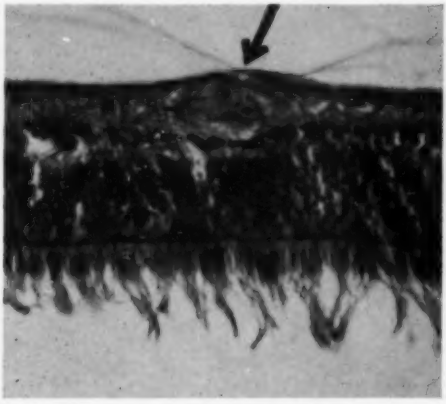

Fig. 3 (Wolter and Phillips). Retina of the posterior pole the inner layers of which show complete atrophy and an occluded artery (arrow). The outer layers are rather well preserved. (Hematoxylin-eosin, photomicrograph.)

The optic nerve was very small and atrophic (fig. 5). The serial cross sections through it revealed the central retinal artery to be completely obstructed by a degenerated mass of curled up layers of tissue. This contained flat cells and obviously represented the hypertrophic and detached intima of this

Fig. 4 (Wolter and Phillips). Flat section through the inner retinal layers which shows the pattern of the brushlike inner processes of the radial fibers of Mueller. (Hortega method, photomicrograph.)

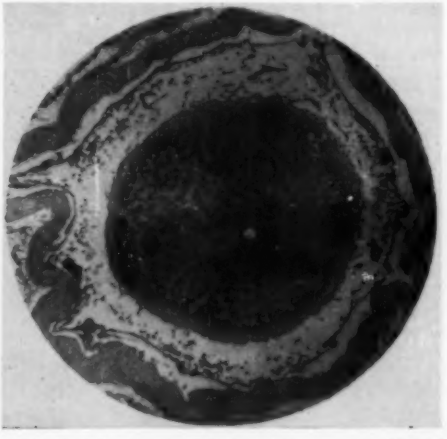

Fig. 5 (Wolter and Phillips). Low-power view of a cross section of the optic nerve which shows the occluded central artery, the open central vein, the atrophy of the nerve, and the loose arachnoidal and dural sheaths. (Hematoxylin-eosin, photomicrograph.)

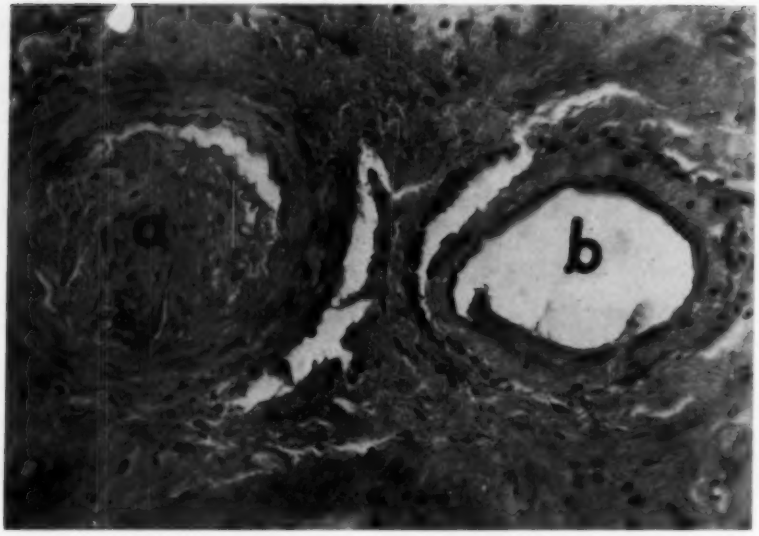

Fig. 6 (Wolter and Phillips). High-power view of the central vessels in the optic nerve. The artery (a) is completely occluded by hypertrophic and detached intima. The vein (b) is open. (Hematoxylin-eosin, photomicrograph.)

artery. This peculiar obstruction of an artery by its own dissected intima is known as "dissecting aneurysm" (figs. 6 and 7). The central retinal vein all through the removed part of the optic nerve was wide open (fig. 6). Some areas of the vein showed an extensive lymphocytic endo- and periphlebitis (figs. 7 and 8). Other areas were without such an infiltration (fig. 6). No epithelioid or giant cells were found in the areas of infiltration around the central retinal vein.

Another interesting finding in the histologic study of this eye was that of two completely occluded posterior ciliary arteries (figs. 9 and 10). These two blood vessels also showed a complete obstruction by curled up hypertrophic intima very similar to that found in the central retinal artery. The other posterior ciliary arteries were open; but they exhibited extensive thickening of their intima.

Comment

In this paper there is no need for a new review of the cases of secondary glaucoma following occlusion of the central retinal artery which were found in the literature and seen earlier by ourselves. This was done in a recent paper on the same subject which has appeared in The Journal.[5] It may be mentioned that two of the five known cases had hypertension. All the known cases, however, had extensive arteriosclerotic changes of the retinal blood vessels.

The time between the arterial occlusion and the occurrence of the secondary glaucoma in the present case was nine weeks. The known cases from the literature show the secondary glaucoma to occur between four and 10 weeks following the occlusive episode (sudden loss of vision).

It is of special interest that the present case, as well as our first case[4] of this entity, exhibited an arterial occlusion not only of the central retinal artery but also of posterior ciliary arteries. This indicates that a regional arterial involvement may be found in such cases and in the future we should always look for the state of these vessels in cases of secondary glaucoma following occlusion of the central retinal artery. It is

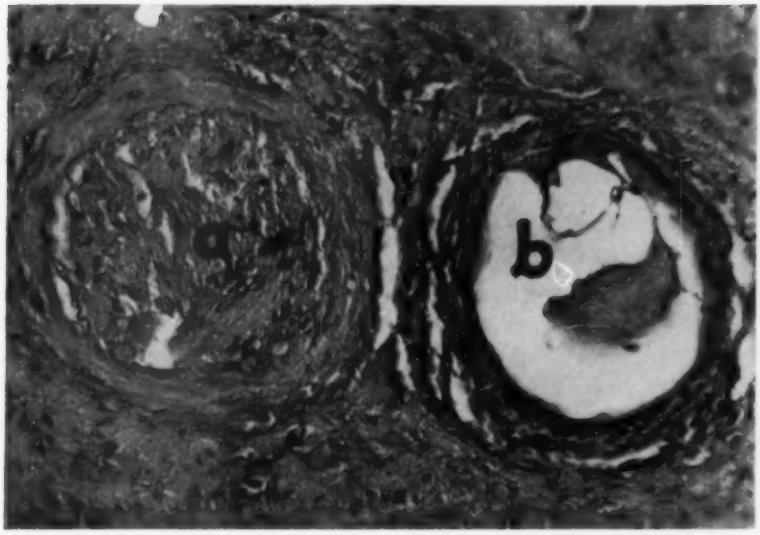

Fig. 7 (Wolter and Phillips). High-power view of another cross section of the optic nerve with the occluded central artery (a). The central vein (b) shows dense lymphocytic endo- and periphlebitis. (Hematoxylin-eosin, photomicrograph.)

very surprising to find that the choroidal arteries in these two cases with involvement of the posterior ciliary arteries showed very little sclerosis.

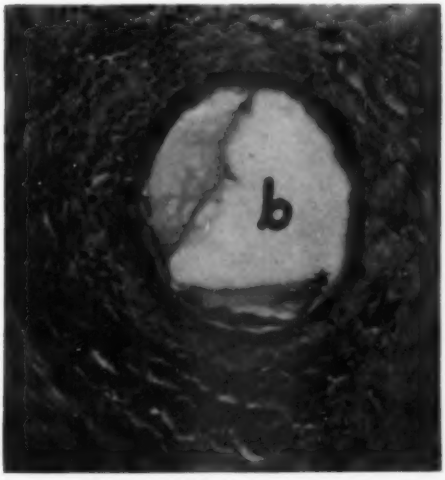

Fig. 8 (Wolter and Phillips). Area of extensive peri- and endophlebitis of the open central vein (b) in the optic nerve. (Hematoxylin-eosin, photomicrograph.)

All three of our cases showed the occlusion of the central retinal artery to be caused by a condition known as "dissecting aneurysm."[7] The question whether the occurrence of this dissecting aneurysm together with the secondary glaucoma in all three of these cases is just coincidence or not is open for further study.

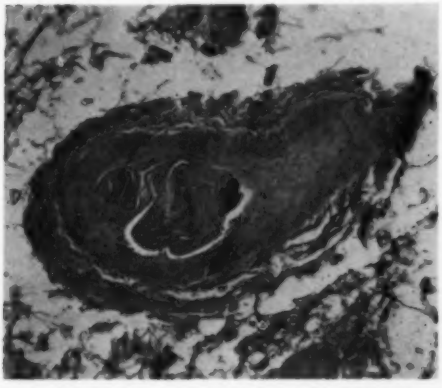

Fig. 9 (Wolter and Phillips). A posterior ciliary artery which is completely occluded. (Hematoxylin-eosin, photomicrograph.)

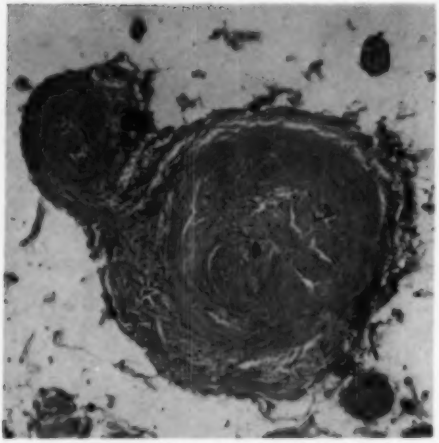

Fig. 10 (Wolter and Phillips). Another ciliary artery which shows complete occlusion. (Hematoxylin-eosin, photomicrograph.)

It would be of great importance to find the cause for the process of proliferation of fibrovascular tissue on the anterior surface of the iris and in the chamber angle which is typically found in secondary glaucoma after arterial occlusion as well as in that following venous occlusion. There can be little doubt that the proliferation of this tissue in the angle represents the direct mechanical cause of the glaucoma.[8] A complete closure of the angle by peripheral anterior synechias on top of the obstruction by fibrovascular tissue occurs often in the late stages of both entities. Both of these changes were well developed in the present case. However, as yet we cannot explain their cause and nature.

A chronic nonspecific endo- and periphlebitis was found in some areas of the central vein. This finding seems to be new in this entity and we have no explanation for it. Our two other cases were without any venous infiltration.

The importance of the present case is not so much that of another occurrence of a relatively little known clinical entity. This case has an exceptionally well-recorded clinical history and a complete histologic study. It should convince those ophthalmologists who still think that the arterial occlusion in these cases of secondary glaucoma occurs secondary to venous obstruction that this entity really exists and that they may see it in their practices. The fact that three eyes with this entity came into our laboratory within the last two years indicates that secondary glaucoma following occlusion of the central retinal artery is not at all rare.

Summary

The history, clinical findings, and histopathology of a case of secondary glaucoma following occlusion of the central retinal artery are described. There can be no doubt that this represents a typical disease entity which occurs in eyes with extensive arteriosclerotic vascular changes. It does not seem to be very rare and is in its late stages easily confused with secondary "hemorrhagic" glaucoma following venous occlusion. The cause and development of the secondary glaucoma following occlusion of the central retinal artery is not yet understood.

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THE PINGUECULA*

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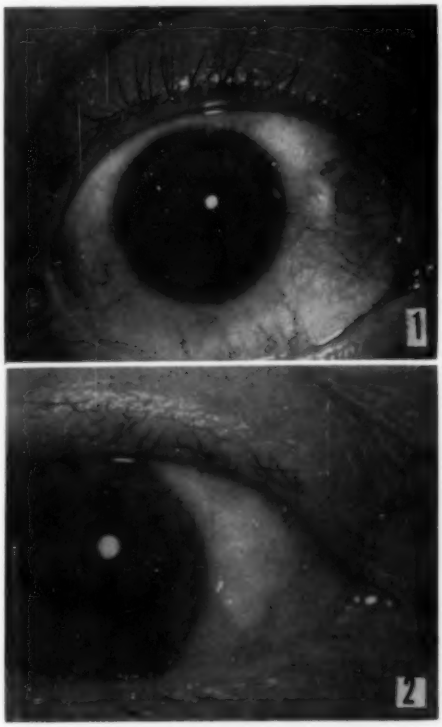
The pinguecula and its sequel, the pterygium, have always intrigued ophthalmologists because of the frequency with which they present themselves clinically. Although the lesions have been studied by many clinicians and pathologists,[1-5] the pathogenesis of these lesions is still not completely understood. As the basis of this report 22 pingueculas and pterygiums that had been excised were studied histologically. In 19 of these the histologic findings were correlated with the clinical appearance of the lesions as independently recorded in color transparencies (table 1).

Pingueculas are usually bilateral and are more often present nasally than temporally (figs. 1 and 2). They consist of yellowish, opaque, soft, slightly elevated tissue varying considerably in size and appearance. In general they are oval or round but tend to be triangular with the base at the limbus. They occupy the interpalpebral area and, depending on the case, usually lie just at the midportion of the corneal margin; the lower portion may extend for some distance along the corneal circumference and for varying distances into the conjunctiva. A pinguecula may be clearly separated by a free area from the cornea but may encroach here to form an early pterygium. In most cases of pterygium the pinguecula appears to be a component of the head of the pterygium and is carried onto the cornea as the pterygium extends.

The area of opacity in the pinguecula consists grossly of single or multiple yellow islands which cover the vessels and fill up the space between the conjunctival epithelium and sclera (fig. 1). In studies of the conjunctival lymphatics with trypan blue[6] it was evident that the lymphatics did not extend into the opaque area. In instances where subconjunctival hemorrhage occurs in the area, or where iritic pericorneal injection is present, the pinguecula usually stands out by its avascularity and relative lack of involvement. At the edges of the pinguecula one usually finds varicosities of the vessels and tortuosity and visible narrowing of the lumen. These vessel changes and absence of lymphatics suggest a degenerative change (figs. 1 and 2).

On histologic examination, the principal lesions occurred in the subepithelial tissues.



Figs. 1 and 2 (Sugar and Kobernick). (1) Close-up photograph of the eye from Case 18, showing a pinguecula with marked prominence of the blood vessels on the nasal side (from Kodachrome). (2) Uncomplicated piguecula from Case 13 (from Kodachrome photograph).

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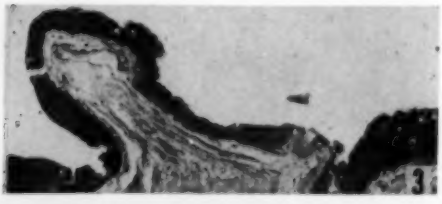
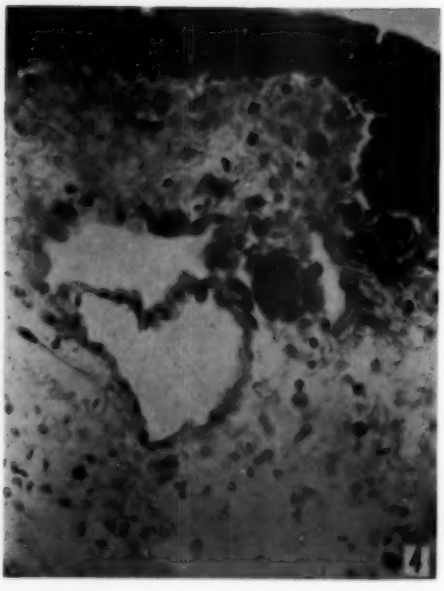
TABLE 1

CORRELATION OF CLINICAL AND PATHOLOGIC FINDINGS

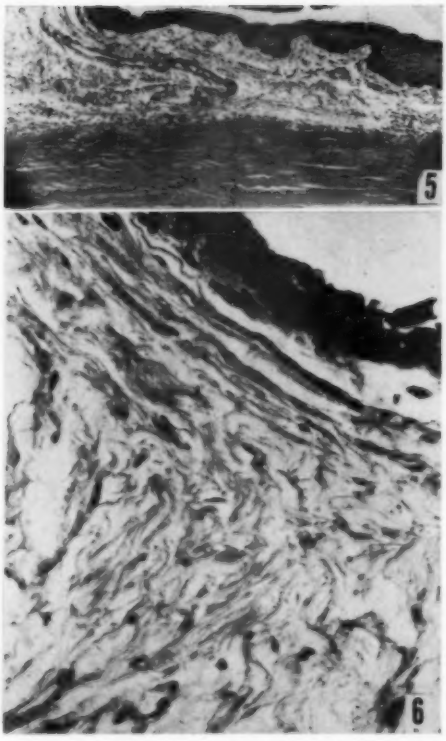
Case No.	Clinical Findings (from color photographs)					Pathologic Findings				
						Epithelium	Connective Tissue			
	Diffuse Yellow Color	Patchy Masses Suggesting Calcif'n	Opacity	Elevation	Conjunctival Irritation	(cells thick)	Hyalinized Collagen	Elastic Hyperplasia	Calcification	Cellular Infiltration
1	+++	+	+++	++	0	5–10	++++	++++	++++	0
2	+	0	+	+	++	8–12	+	+	++++	0
3						5–10	++	++++	0	0
4						5–8	+++	++	+++	0
5	+++	+	+++	++	++	10–15	++	++	+++	+ Focal Acute
6						8–15	+	++	0	0
7	0	0	++	+	+	5–8	+++	+++	+++	0
8	0	0	+++	+	+	5–8	+	+	0	0
9	+	+	++	++	+	8–40	++	++++	++	++ Lymphs Subepithelial
10	0	0	+	0	+	3–8	+	++++	0	0
11	0	0	++	++	+	5–8	++	++	0	
12	+++	+	++	+	+	5–20	++	+++	0	Moderate vascularization
13	0	0	+++	+++	0	8–15	++	++	0	0
14						5–6	+++	++++	0	0
15	0	0	+++	+	+	degeneration	++	++++	0	+ In elastic tissue
16	0	0	++	+	0	degeneration	++	++++	0	+++ Perivascular
17						5–20	+++	++	++	?
18	0	0	+++	++	+	8–20	+	++++	0	+
19	—	—	+++	+	0	5–8	+	+++	0	0





The epithelial layers themselves showed no significant alteration (fig. 7). Some cases had a slight increase in the number of squamous cells, from five to eight, but, in general, the number and configuration of the superficial conjunctival epithelium was normal (figs. 3, 4, 5, 6, 8, and 9). The layers immediately subjacent to the epithelium showed marked hyalinization of collagen (fig. 7). This was succeeded rapidly by a hyperplasia and distinct increase in elastic tissue fibers (figs. 9 and 10), often increasing the thickness of the subepithelial tissues 10 to 12 times the width of the epithelium and 20 times normal (fig. 7). In the elastic tissue layers, clumps of irregularly degenerated material, taking the elastic stain but also showing evidence of basophilia—suggesting calcification—were often found (figs. 7 and 8). These were also deposited within the hyaline

Figs. 3 and 4 (Sugar and Kobernick). (3) Normal conjunctiva excised during a cataract operation. (Hematein, phloxine, and saffron; ×90). (4) Normal conjunctiva from same case as shown in Figure 3, showing the delicacy of the tissue and a large lymphatic channel, as well as the usual thickness of the conjunctival squamous epithelium. (Hematein, phloxine, and saffron, ×370).



Figs. 5 and 6 (Sugar and Kobernick). (5) Normal conjunctiva and sclera from an excised eye. (Hematein, phloxine, and saffron; ×90). (6) Normal conjunctival epithelium and subconjunctival connective tissue of the visceral conjunctiva. Note the extreme delicacy of the connective tissue pattern beneath the thin epithelium. (From same case as in Figure 5. Hematein, phloxine, and saffron; ×370).

layer. Their continuity with the hyperplastic elastic tissue was evident in some cases. Deep in the hyperplastic elastica the subconjunctival connective tissue resumed its normal vascularity and lymphatic channel content (fig. 7). A summary of the severity of the changes can be found in Table 1.

The attempt to predict the histologic picture from clinical photographs, in those cases where these were available, were unsuccessful (table 1).

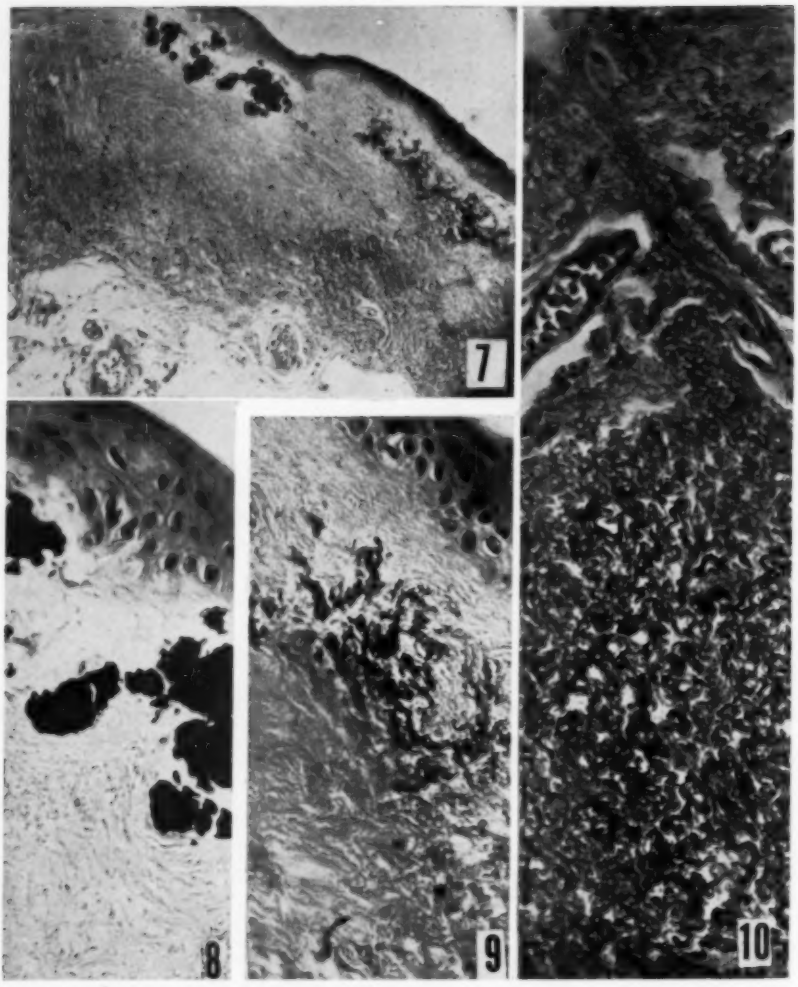
The histologic structure of pinguecula is thus characterized by four processes: (1) hypertrophy and hyperplasia of elastic fibers, (2) hyaline degeneration of collagen fibers, (3) deposition of amorphous hyaline substance, and (4) the formation of concretions. The histologic picture was first described by Fuchs and later corroborated by Hubner.[5] The changes are particularly striking in contrast with the normal structure of the conjunctiva at the limbus where the tissue consists of very loose connective tissue arranged around a lymphatic meshwork with only an occasional elastic fiber in evidence (fig. 4).

Discussion

The degenerative nature of pingueculas has been inferred by Hinnen[7] who found an increasing incidence of pingueculas with aging. Of 462 individuals with normal eyes, he found that in 48 patients over 70 years of age, only two had no pinguecula. Some few were present in those between 10 and 20 years of age.

The principal characteristic of the pinguecula histologically is the hyalinization of the subepithelial tissue and the remarkable hyperelastosis. The normal conjunctiva contains practically no elastic tissue and this elastosis is the principal component of the lesion. The hyalinization of the superficial tissue and ultimate calcification must be considered to be derived from the increased elastica that undergoes degeneration. It is remarkable too that in uncomplicated pinguecula there is a very small inflammatory component. Little is known about elastosis. Authorities on skin pathology cite only "hyperelastosis cutis" as an instance of elastic tissue disorders. However, it should be noted that this increase in elastic tissue may be found adjacent to subacute inflammations or ulcerative inflammations of the mouth and of the skin. It is also noteworthy that in the so-called "collagen diseases" elastic tissue is as much affected as is collagen by the fundamental lesion.[8]

To understand the pathogenesis of the pinguecula one must consider conjunctival anatomy and its relation to the physiology of lid blinking. When the lid blink reflex occurs,



Figs. 7, 8, 9, and 10 (Sugar and Kobernick). (7) Excised pinguecula from Case 1. Note the thickening of the entire conjunctiva, the comparative normality of the superficial squamous layer, the hyalinization of the subepithelial connective tissue, calcium deposition left center, and right the fragmentation of the elastica continuous with the fragmentation of the elastica in the deepest layers. (Hematein, phloxine, and saffron; ×90). (8) A portion of the subepithelial layer in which there is focal calcification represented by the black masses. (Hematein, phloxine, and saffron; ×270). (9) A portion of the conjunctiva beneath the epithelium in which there is marked fragmentation of elastica. It shows the hyalinization of the subepithelial connective tissue. (Hematein, phloxine, and saffron; ×270). (10) Same case showing the deep layers of elastic tissue with fragmentation, with increase in elastica, reduplication and fragmentation of elastic tissue fibers (Weigert's elastic stain).

the interpalpebral conjunctiva is squeezed between the lids each time. The portion of conjunctiva adjacent to the limbus is fixed to the underlying sclera while the remaining portion is movable. Thus with the formation of a pinguecula the involved interpalpebral area, being partly fixed, receives the effect of the additional conjunctival tissue

squeezed into the area from above and below by the lids. Observation suggests that the nasal interpalpebral area is more firmly pressed by the lids than the temporal portion because of the greater curvature of the orbicularis nasally. This may explain the greater frequency of pinguecula nasally.

In some instances the patient's attention is called to a pinguecula when it begins to show signs of irritation. In these cases the epithelial surface shows increased whiteness in its center. Staining with fluorescein is usually present. Histologically there is subepithelial round-cell infiltration in addition to the degenerative tissue usually present in a pinguecula. It is doubtful that this inflammation is a major factor in the causation of pinguecula; however, it probably contributes its share. The simplest method of treatment is removal of the total pinguecula area including the area of inflammation down to the scleral surface.

The pathogenesis of the lesion may be considered as originating with some stimulus, such as the physical motion of the lids against the conjunctiva, along with age changes, allergy, and possibly mild inflammatory factors. This could imitate a degenerative and reparative reaction, manifested by elastic tissue formation and collagen-elastic degeneration. The swelling associated with the latter process causes elevation of the tissue. The process tends to be a continuing cycle since the elevated tissue tends to more pressure by the lids and further reparative-degenerative reaction. In those cases where the volume of the pinguecula is enough to involve the edge of Bowman's membrane, which seems to act as a barrier, it is suggested that pterygium has its beginning. As the pterygium "grows," the pinguecula is carried onto the cornea at its head.

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BLUE SCLEROTICS SYNDROME SIMULATING BUPHTHALMOS*

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A newborn infant with cloudy corneas does not necessarily have congenital glaucoma. Birth injuries may produce corneal opacities due to ruptures in Descemet's membrane. Hurler's disease and, rarely, luetic interstitial keratitis may present with corneal infiltrates in the newborn. If, in addition to cloudy corneas, the patient has blue scleras, he may be manifesting ocular complications of the osteogenesis imperfecta syndrome.

Osteogenesis imperfecta is a genetically transmitted disease characterized by defective mesodermal structures throughout the body. Blue scleras are the most consistently present sign of the syndrome. Fragile bones and lax joint capsules leading to frequent fractures and dislocations are present in many; in addition, otosclerotic deafness develops in about 50 percent during the second

*Presented at the Massachusetts Eye and Ear Infirmary Alumni Meeting, April, 1958.

Fig. 1 (Tucker). J. B., right eye at age of three weeks, showing cloudy, bulging cornea.

or third decade of life.[1] Some are found to have congenital heart disease.

The structural defect reputed to be responsible for transparency of the sclera, namely a deficiency of collagen and retention of immature reticulum fibers, is shared by the cornea.[2] Megalocornea, myopia, keratoconus, and corneal infiltrates develop in some patients with the disease.[3-6] It has been suggested that these corneal changes may also be secondary to the collagen deficiency.[2]

Case report

There have been no previous reports of corneal changes so severe in the newborn as to confuse the diagnosis with buphthalmos. The following is a report of such a patient.

J. B. was the second of two children born to parents who were first cousins. His delivery was by caesarian section after a 24-hour ineffective labor complicated by the development of a uterine contraction band. In his second day of life he began to vomit; and on the third day he developed hematemesis so severe as to require transfusions. A barium swallow and gastrointestinal series did not reveal the source of the bleeding. On general examination at this time he was noticed to have a heart murmur, bilateral congenitally dislocated hips, unusually long, slender fingers, an umbilical hernia, and cloudy corneas of both eyes.

With a presumptive diagnosis of buphthalmos, the patient was transferred at the age of three weeks to the Massachusetts Eye and Ear Infirmary. Examination of the eyes showed cloudy, bulging corneas and very deep anterior chambers (fig. 1). Corneal diameters were within normal limits and no breaks in Descemet's membrane were seen. There were good red reflexes, but fundus details could not be discerned. Tensions were found to be: O.D., 16; O.S., 20 mm. Hg (Schiøtz, 1948 calibration). The consensus was that the patient had congenital glaucoma and surgery was contemplated. But because of persistently normal tensions and the uncertainty of diagnosis he was discharged untreated.

Two months later the infant was readmitted for tonometry and fundus examination under general anesthesia because the right cornea now had developed a break in Descemet's membrane. Since the last examination, however, both corneas had cleared considerably in the periphery. Tensions were still normal: O.D., 20; O.S., 20 mm. Hg. (Schiøtz). The discs, macular areas, and central retinas appeared normal. The peripheral retinas showed some peculiar scarring and bluish discoloration. He was again discharged without further elucidation of the corneal condition.

After that he was seen in the eye clinic every few months, and was noted to have apparently normal tensions, progressive clearing of both corneas, persistently deep anterior chambers, and normal appearing fundi. On two occasions a blue tint to the scleras was noted. At the age of 18 months the child was recognized to have a pendular nystagmus. In addition, a diagnosis of patent ductus arteriosus was made at the Children's Medical Center.

On a routine clinic visit, when the patient was nearly three years of age, his four-year-old sister was brought along. The diagnosis was now evident. Both children had blue scleras (figs. 2 and 3) and other evidence of the osteogenesis imperfecta syndrome. The more subtle characteristic physical findings of short, broad heads and long, slender, hyperextensible fingers were present in both. The sister had been in excellent health except for a fractured clavicle sustained in a fall from a chair at the age of three years.

Slitlamp examination revealed uniformly thin

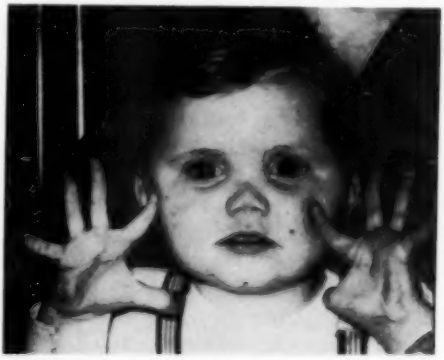

Fig. 2 (Tucker). J. B., at age of three years, showing blue scleras, brachycephaly, and arachnodactyly.

Fig. 3 (Tucker). M. B., at age of four years, showing blue scleras, brachycephaly, and arachnodactyly.

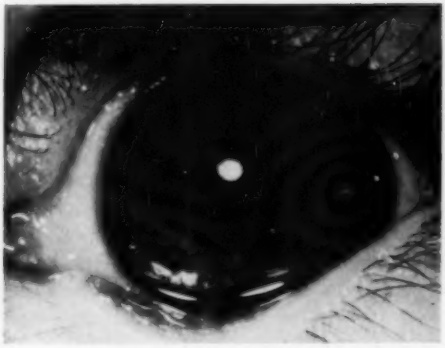

Fig. 5 (Tucker). J. B., left eye at the age of three years, showing blue sclera.

corneas in both children. J. B. (figs. 4 and 5) showed a faint superficial central stromal nebula in the right eye. His sister had no corneal opacities. Both children were found to have high myopic regular astigmatism on retinoscopy. J. B.'s corrected vision, wearing a −10.0D. sph. ⊃ +5.0D. cyl. ax. 90°, O.U., was 20/200 in both eyes. It did not improve after wearing glasses for six months, presumably because of the nystagmus. His sister's vision, after wearing −5.5D. sph. ⊃ +4.0D. cyl. ax. 100°, O.D., and −5.5D. sph. ⊃ +4.0D. cyl. ax. 80°, O.S., improved from 20/200, O.U., without correction to 20/70, O.U., with correction.

Comment

In retrospect, it appears that the patient reported here had many signs and symptoms of his disease practically from the day of birth. Although spontaneous gastro-intestinal hemorrhage is not characteristic of the osteogenesis imperfecta syndrome, excessive lacerability of blood vessels has been noted.[7] The trauma of prolonged vomiting may have been sufficient to cause the hematemesis two days after birth. In the neonatal period he was found to have a heart murmur, dislocated hips, an umbilical hernia and slender fingers. All are part of the syndrome. In addition, nystagmus such as noted in this patient has been reported and ascribed to labyrinthine disturbances.[8] The most constant feature of the disease, blue scleras, went long unrecognized.

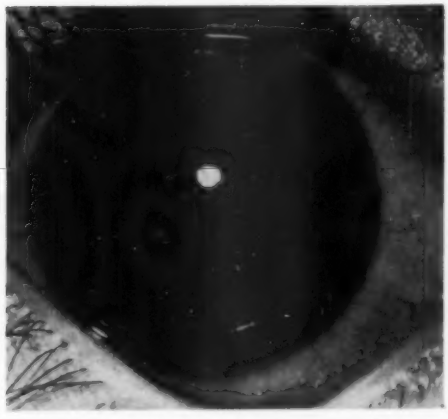

Fig. 4 (Tucker). J. B., right eye at age of three years, showing blue sclera and corneal nebula.

The reason for bilateral corneal cloudiness at birth in this patient is not obvious. Spontaneous tears in Descemet's membrane in both eyes seems a likely possibility, since one such lesion was noted in the right eye at the age of three months. The long, obstructed labor may have been adequate stress to produce tears in these pathologic corneas. However, no breaks in Descemet's membrane were recognized during the patient's initial hospitalization.

Blue scleras have a number of different etiologies. In many normal newborns there is a physiologic blue tint which gradually fades in later months as the scleras mature. This makes the differentiation of normal from pathologically blue scleras more diffi-

cult in infants than in older children and adults.

In patients with buphthalmos, the picture is further complicated by the fact that stretching of the ocular tissues thins the sclera and cornea, allowing uveal pigment to show through. Such patients may have enlarged eyes with cloudy corneas and blue scleras.

When these etiologies are thought unlikely, hereditary diseases of connective tissue must be considered in the differential diagnosis of blue scleras, with or without associated corneal changes. Such findings have been noted in the osteogenesis imperfecta syndrome (discussed in the introductory paragraphs) and in the Ehlers-Danlos and Marfan's syndromes.

The Ehlers-Danlos syndrome, characterized by hyperelastic, fragile skin, easy bruisability, and hyperextensible joints, has been at times associated with such ocular abnormalities as blue scleras, microcornea and glaucoma, dislocated lens, and retinal hemorrhages.[9]

Marfan's syndrome is characterized by exceptionally long arms and legs, a long, narrow head, and aortic insufficiency or dissecting aneurysm of the aorta. The majority of those affected have dislocated lenses. In addition, blue scleras,[10] keratoconus, and megalocornea have been found in some.

Fortunately, the patient reported here was not operated on for "glaucoma." It is apparent that blue scleras must be looked for whenever cloudy corneas are discovered in an infant. And when blue scleras are found in the presence of normal-sized eyes with normal tensions, it should be suspected that a systemic disease of connective tissue is the underlying cause. Examination of the patient and members of his family for the characteristic associated findings will make the diagnosis in such cases and avert the possibility of inappropriate therapy.

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A NEW AND OBJECTIVE METHOD FOR MEASURING OCULAR TORSION*

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INTRODUCTION

There is a need for a reliable method of measuring rotation of the eye whether we are concerned with ocular movements per se or with their usefulness as a reflex indicator of another function. To this end a number of methods have been devised utilizing artificial[1] or natural[2] landmarks and after-images.[3] For various reasons none was found suitable for our purpose, which led to the development of the procedure now to be described. It consists essentially of photographing artificial markers on eye and head before, during, and after a test procedure and measuring, in the frontal plane, the change in angle between a reference line in the eye (eye line) and a reference line on the head (head line).

PROCEDURE

The marks on the head consisted of a 5.0 by 5.0 mm. black square on a white plastic tape, placed near the outer canthus of each eye (fig. 1). The marks on the eye were black silk sutures. Usually, the master eye was used. The bulbar conjunctiva was anesthetized with 0.5-percent Pontocaine. Additionally, two-percent Procaine was infiltrated subconjunctivally between the limbus and the inner canthus. The sutures were placed subconjunctivally near the limbus on either side of the cornea along an imaginary horizontal line passing through the anterior pole of the eye. A knot was tied at the central end of each suture and then pulled snug; another knot was tied at the lateral end to prevent the suture from slipping, and the free ends were clipped close to the knots (fig. 1). Sutures were left in place as long as four hours. During this period a drop of 0.5-percent Pontocaine was used if necessary. After the stitches were removed, a small area of redness marked the site, but this usually cleared by the next day. In a large series of trials the only complications were small subconjunctival hemorrhages. Several subjects were used repeatedly.

Photographs were taken with a camera equipped with a remote firing device, and either a shutter-synchronized electronic flash firing at 1/1,000 sec., or a flash bulb with 1/200 sec. shutter. The camera was mounted in front of the subject, and the relative position of the subject's head and camera was maintained throughout. A dim red light near the lens served as a fixation point, and the subject was requested to fixate the light prior to film exposure. The 4.0 by 5.0 inch negatives were enlarged to one and one-half times life size.

The photograph was placed on a sheet of paper to make the measurement, and photograph and paper were tacked on a drawing board. With the use of a hand lens and a fine stylet, pin pricks were made at one emergent point of each suture, at one corner of each black square, and at the inner canthus of each eye in the picture. On the reverse side of the photograph, fine lines were drawn connecting paired points. The same was done on the underlying paper. The

Fig. 1 (Graybiel and Woellner). Drawing of subject's eyes, with sutures and outer canthal landmarks in place.

*From the U. S. Naval School of Aviation Medicine, U. S. Naval Aviation Medical Center. Opinions and conclusions contained in this report are those of the authors and do not necessarily reflect the views or endorsement of the Navy Department.

TABLE 1

ANGLES FORMED BY INTERSECTION OF EYE-LINE WITH INNER CANTHUS LINE (IC) AND WITH OUTER CANTHUS LINE (OC) AS MEASURED BY TWO OBSERVERS FROM 20 PHOTOGRAPHS OBTAINED FROM ONE SUBJECT SEATED IN THE DARK

Photo No.	Eye Inner Canthus Angles		Eye Outer Canthus Angles	
	Observer 1 Degrees	Observer 2 Degrees	Observer 1 Degrees	Observer 2 Degrees
1	13.1	12.6	11.5	10.8
2	13.4	12.4	11.2	10.6
3	13.3	12.4	11.2	10.5
4	13.4	12.6	10.9	10.6
5	13.2	12.6	11.0	10.6
6	13.0	13.4	10.8	11.0
7	13.7	13.6	10.8	10.0
8	12.4	12.4	10.4	10.0
9	13.7	13.4	11.2	10.6
10	13.3	13.0	11.0	10.6
11	13.8	12.7	11.7	10.6
12	13.8	13.6	11.5	11.0
13	14.0	13.8	11.4	11.1
14	14.5	14.1	11.7	11.6
15	13.7	13.4	11.5	11.0
16	14.2	14.0	11.8	11.6
17	14.3	14.0	11.3	11.4
18	14.2	13.6	11.6	11.0
19	13.9	13.4	11.2	11.0
20	13.9	13.3	11.5	10.8

angles formed by the intersection of the "variable" eye line with the "fixed" reference lines were measured with a protractor. These formed the data from which all other measurements were derived.

DISCUSSION

The reliability of the procedure is shown from the measurements, made by two observers working independently, which are summarized in Table 1 and Figures 2 and 3. The measurements in Table 1 were made from 20 consecutive photographs of a subject seated upright in a dark room. The first six were taken at approximately 100-second intervals with the subject stationary. The remaining 14 were taken over a period of one hour, the subject having moved about or changed position between exposures.

One factor affecting reliability was the accuracy with which the angles were measured. Inasmuch as the photograph and the

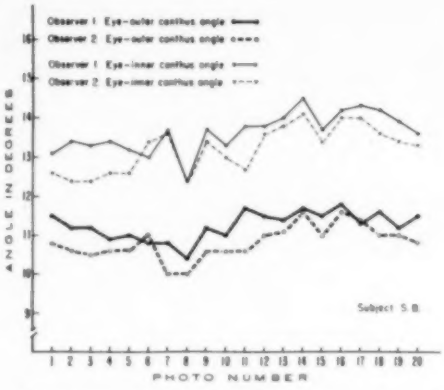

Fig. 2 (Graybiel and Woellner). Comparison of measurements made by two observers from duplicate prints of 20 photographs.

underlying paper were pierced simultaneously by the stylet, the resulting measurements on each should be the same. The mean difference between the measurement of the angles on film and paper in the 20 instances was 0.14 degrees for one observer and 0.11 degrees for the other. This is satisfactory for most purposes. Specially devised equipment would be needed to increase the reliability here.

A second factor was the consistency with which the observer could pierce refer-

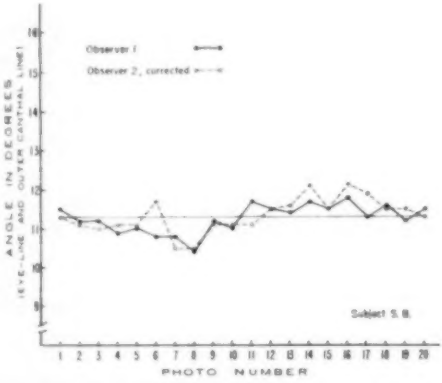

Fig. 3 (Graybiel and Woellner). Comparison of measurements by two observers, using identical prints from 20 photographs.

ence points in a series of photographs. This was tested by two observers by making measurements on five prints from the same negative. The maximum variation was 0.3 degrees in determining the angles.

A third factor was the stability of the fixed reference line on the head. Two lines were used; one connected the inner canthi, and the other connected markers near the outer canthi. The inner canthi had the advantage that they were firmly attached to underlying tissue and were in nearly the same plane as the eye sutures. They had the disadvantage in that it was difficult to pierce identical points from one photograph to another. The outer canthal markers had the advantage that identical points on the black squares were easy to locate; these markers had the disadvantage, however, that they were located on movable skin and were posterior to the plane of the eye. An indication of the stability of the two lines was the consistency of the angle between them. The variation was less than 0.2 degrees in 15 of the 20 measurements made by one observer and 11 of the 20 made by the other observer.

There still remains for consideration the inter-observer variation seen throughout the series of 20 trials. It is seen from Figure 2 that the two observers obtained different values in measuring the angles. This is unimportant if the difference is constant. Observer 2 obtained smaller values than Observer 1 except in two instances. In the measurement of the angles utilizing the inner canthal line the mean difference was −0.43 degrees and for the outer canthal line, −0.48 degrees. Undoubtedly, slightly different points were used in determining the emergence of the sutures. When these amounts were added to the values obtained by Observer 2 the agreement between the two observers was good. Figure 3 shows correspondence of corrected and uncorrected outer canthal series. There was a difference of 0.2 degrees or less in 12 of the 20 measurements where the inner canthal line was used and in 13 of the 20 where the outer canthal line was used. The greatest difference was only 0.7 degrees.

In view of the high reliability of the individual measurements and the remarkable consistency between the corrected values obtained by the two observers, it is highly probable that the major changes shown in Figure 2 represent spontaneous variations in the position of the eye. Such variations have been regularly observed in the subjects tested. Consequently, control measurements were made before and after each experimental trial, and the average of the two was used as the baseline value. The measurement made during the experimental trial was subtracted from this baseline value, and the difference was the amount of torsion during the trial. Subsequent reports will reveal some of the ways in which this method may be exploited.

Summary

A procedure was devised to measure accurately torsion of the eye. This procedure was as follows: The bulbar conjunctiva of the master eye was anesthetized, and sutures were placed to the left and right of the cornea in line with the anterior pole of the eye. The conjunctival entry of each suture constituted ocular reference points. Two pairs of reference points on the head were used. The subject was photographed before, during, and after the experimental trial. Lines were drawn on the photograph connecting paired points, and the angles between the eye line and the fixed reference lines were determined. In serial photographs differences in these angles were due to rotation of the eye. The procedure is objective and highly reliable if carefully carried out.

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We are grateful to Dr. James L. Fuelling and Dr. Thomas W. Turner for applying the sutures.

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SURGERY OF PTOSIS ASSOCIATED WITH JAW-WINKING*

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Jaw-winking in ptosis patients was first described in 1883 by Marcus Gunn.[1] Its incidence in patients with drooped lids is two to four percent. Berke[2] found seven cases with jaw-winking in 200 consecutive ptosis cases, while Spaeth[3] diagnosed two cases in 100. In a previous paper, I[4] reported, in a series of 142 ptosis patients, six cases with jaw-winking.

All six patients had unilateral congenital ptosis in which there was a retraction of the lid upon opening the jaw. Three were males and three were females. Except in one child, the left eye only was affected.

Surgery was performed in four patients who had considerable ptosis and jaw-winking; surgery was deferred in one because the child was just two years of age; while in another (fig. 1), surgery was not advised because the ptosis measured barely a millimeter and the jaw-winking was hardly noticeable, except during exaggerated chewing movements.

As shown in Table 1, different types of surgery were performed in these four Childrens Hospital cases, as in one case done elsewhere. While it is true that the number of these surgical cases is inadequate for statistical purposes, a few observations might serve to guide the management of the Marcus Gunn phenomenon. In all five cases, the levator muscle was grossly normal in size and in elasticity. When the levator muscle was simply shortened (fig. 2), jaw-winking persisted, although somewhat less during the first few days after surgery, possibly due to transient surgical trauma on the levator muscle. On the other hand, when tenectomy of the levator was done (fig. 3) and later the orbicularis and frontalis muscles were utilized (Reese's method[5]), jaw-winking was completely disrupted, and no significant increase in amblyopia was noted. However, when the procedure was done in one operation, utilizing tantalum wire as a sling to

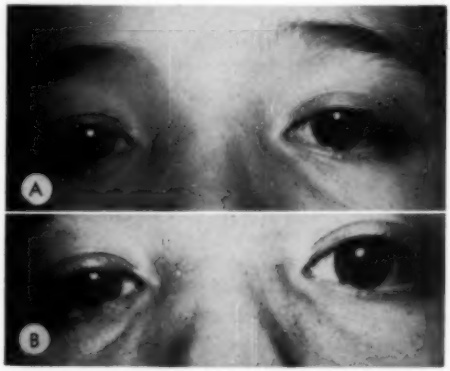

Fig. 1 (Carbajal). An example of mild jaw-winking, requiring no surgery. (A) Mouth shut. (B) Mouth open.

* From the Los Angeles Childrens Hospital and the University of Southern California School of Medicine.

TABLE 1

SURGICAL MANAGEMENT OF THE MARCUS GUNN PHENOMENON
(Five cases*)

Case No.	Race	Sex	Age at Surgery (yr.)	Eye	Type of Surgery	Follow-Up			
						After First Month		Six Months or Later	
						Jaw-winking	Ptosis	Jaw-winking	Ptosis
1	W	M	5	O.S.	Levator tucking	Less	Less	Complete recurrence	Complete recurrence
2	W	M	9	O.S.	Blascovics	Less	Less	Complete recurrence	Partial recurrence
3	Mexican	F	6	O.S.	Two stages: tenectomy and Reese	Gone	Gone	Gone for 23 mo.	Partial recurrence
4	Mexican	F	9	O.S.	One stage: tenectomy and Friedenwald-Guyton	Gone	Gone	Gone for 8 mo.	Slight recurrence
5†	W	M	6	O.D.	One stage: tenectomy and Friedenwald-Guyton	Gone	Gone	No recurrence yet	

* The Marcus Gunn phenomenon had been present since birth in all five cases.
† Surgery was done at Saint Vincent's Hospital.

the frontalis (fig. 4), there was slight and gradual recurrence of jaw-winking in one patient, while in another (fig. 5) the result to date is satisfactory.

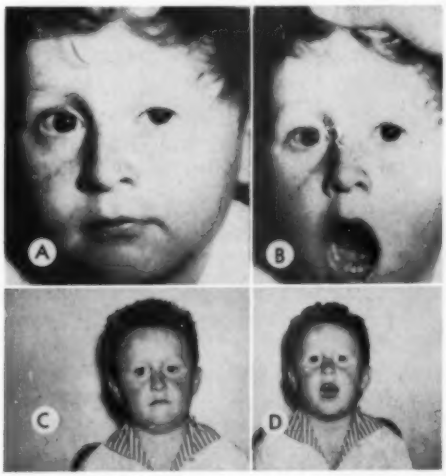

Fig. 2 (Carbajal). An example of moderate jaw-winking, in which levator tucking was done. (A and B) Preoperative pictures. (C and D) twenty-eight months after surgery. Note the retraction of the left upper lid when the mouth is opened wide.

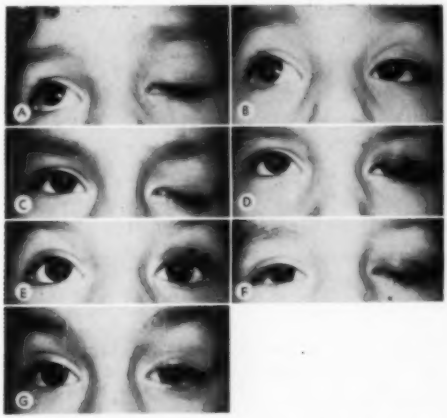

Fig. 3 (Carbajal). An example of marked jaw-winking, in which a two-staged procedure was done. (A and B) Preoperative pictures, mouth shut and mouth open. (C) Three weeks after first stage—tenectomy of levator muscle. (D and E) Three weeks after second stage—Reese's orbicularis-frontalis sling, without and with frontalis action. (F and G) Six months after second stage, without and with frontalis action. Note the slackening of the "sling," resulting in marked ptosis. The jaw-winking reflex was completely disrupted for 23 months and then gradually reappeared, although not as marked as before surgery.

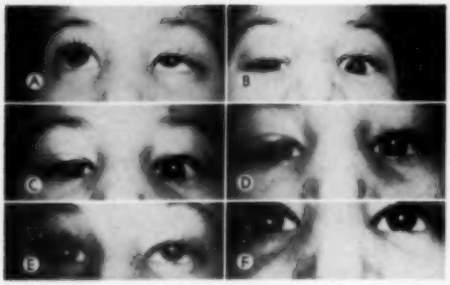

Fig. 4 (Carbajal). Another example of marked jaw-winking. In this case, tenectomy of the levator muscle and the Friedenwald-Guyton technique were done at one operation. (A and B) Preoperative pictures, mouth shut and mouth open. (C and D) Ten days after surgery, mouth shut and mouth open. It is evident here that the mandibulopalpebral synkinesia has been successfully broken. (E and F) Twenty-seven months after surgery. There is only slight recurrence of jaw-winking.

The chief trouble in the utilization of an orbicularis tongue as a sling attached to the frontalis (Reese's method) was the tendency for the upward pull to slacken with the passing of time (fig. 3). Moreover, when the frontalis was not active, ptosis was quite evident. In one (fig. 4) of the two patients in which the Friedenwald-Guyton technique[6] (utilizing tantalum) was performed, photophobia developed, lasting for many months even after corneal staining was no longer demonstrable. This was largely due to impairment of the blinking reflex and some exposure keratitis postoperatively, which cleared away rather slowly.

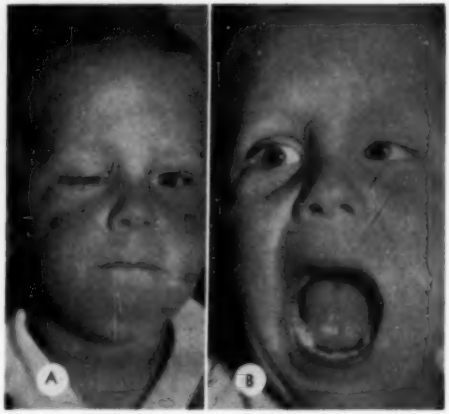

Fig. 5 (Carbajal). Another example of marked jaw-winking, this time in the right eye. (A) Mouth shut. (B) Mouth open. Tenectomy of the levator muscle and the Friedenwald-Guyton procedure were performed at one operation.

It is noteworthy that, with the exception of one, all of these patients presented amblyopia in the ptotic eye and muscle imbalance, four having hypotropia on the affected side (table 2).

Comment

There is little in the literature regarding the management of a case showing the Marcus Gunn phenomenon, in comparison to the lengthy discussions on the etiology or mechanism of production of the phenomenon. Various authors have suggested certain surgical procedures to interrupt the mandib-

TABLE 2

Incidence of heterotropia and amblyopia in five patients showing the Marcus Gunn phenomenon

Case No.	Visual Acuity		Muscle Status
	O.D.	O.S.	
1	20/20	20/20	Orthophoria
2	20/15	20/70	Hypotropia, O.S., weak left superior rectus
3	20/30	20/200	Hypotropia, O.S., weak left superior rectus
4*	20/30	20/200	Hypotropia, O.S. (slight), esotropia, 20°
5†	20/60	20/20	Hypotropia, O.D., weak right superior rectus, weak right lateral rectus

* Recession of left medial rectus and resection of left inferior rectus was done with good result.
† Recession of right medial rectus and fenestrating tenotomy of right inferior rectus gave a good cosmetic result.

ulopalpebral synkinesia and to correct the ptosis. To mention a few, Spaeth[7] advocates dissecting the entire levator from the tarsal plate and superior cul-de-sac, thus creating a complete paralytic ptosis, which is later corrected by a sling procedure. Likewise, Fox[8] advises that, even though the levator is intact, the ptosis must be made complete by levator recession or tenectomy. He does not specify, however, which procedure is best for the correction of the complete ptosis thus produced. Callahan[9] suggests tenectomy of a small section across the width of the levator just above the upper edge of the tarsus; then the ptosis is corrected by attaching the lid to the frontalis or to the superior rectus. Berke's[8] technique has been suggested but, like the Blascovics method,[10] is likely to fail in correcting the jaw-winking, since in these two procedures, the levator muscle still continues to elevate the lid.

Foreign authors (Winkelman[11] and Raimondo[12]) have recently reported some success in surgery for ptosis and jaw-winking. The former corrected the ptosis with the Friedenwald-Guyton suture, while the latter used the method of Cattaneo, combined with the method of Angelucci as modified by Bietti.

The Marcus Gunn phenomenon, Kanter[13] believes, is not so rare as is generally assumed. However, judging from the number of cases observed in Los Angeles Childrens Hospital, where a large volume of eye patients is seen, it is a rare eye condition. Although no bilateral cases have been seen in this hospital, the phenomenon may be observed in both eyes, according to Amat[14] and Kanter.[13]

Simpson,[15] after thoroughly reviewing the concepts on the etiology of Marcus Gunn phenomenon up to 1955, states that abnormal nervous connections are favored by most authors. The malconnections may be cortical, subcortical, internuclear, or infranuclear (peripheral). Ascher[16] reported a case of Marcus Gunn phenomenon during the patient's recovery from a unilateral third-nerve palsy.* It would not be surprising if Simpson's case[15] had a latent jaw-winking before ptosis surgery, which became manifest when the Blascovics technique was performed. This was possibly due to tightening of the levator and Müller's muscles, thus enhancing the retraction of the lid.

Another etiologic hypothesis is that of Duke-Elder,[18] who considers a physiologic explanation more acceptable—that is, there is irritation of normally dormant connections, or release of inhibition, or spread of impulses by irradiation.

A third explanation is that there is a release or irritation of an ancient phylogenetic reflex. This is supported by Ascher,[16] Brain,[19] and Wartenberg.[20] Recently, a pseudo-ophthalmoplegic explanation of the Marcus Gunn syndrome was advanced by Haynes.[21] In discussing the case, Nugent[22] cited two cases of head injury resulting from auto accidents.

It appears that a neuromuscular connection is responsible for the mandibulopalpebral synkinesia. Milano[23] tried to interrupt the synkinetic arch by injecting substances which have a paralyzing effect on the sympathetic system and on the inferior motor centers, but failed.

The observation that almost all of the Marcus Gunn patients in this series, as well as that of Spaeth,[3] had muscle imbalance, usually hypotropia of the ptotic eye, brings up many considerations. When the mouth is opened in these patients, there is no upshooting of the eyeball to correspond with the lid retraction. This indicates that there is some dissociation of the innervation of the levator and superior rectus muscles. This is difficult to understand since embryologically and anatomically the two muscles are closely related. But one should be reminded at this point that ptosis is not always associated with superior rectus underaction.

That there is latent power in the levator is

* Hartman,[17] on the contrary, has pictures of a case which developed reversed Marcus Gunn phenomenon as a seventh-nerve palsy cleared.

proved by the fact that, when the jaw is opened, there is rapid elevation or retraction of the eyelid. This is also borne out by the finding of normal levator muscles during surgery in all five cases. In his series, Spaeth[5] found the levator muscles to be of normal structure, while the other muscles (superior rectus in three, external rectus in two, inferior oblique in two) showed a pathologic condition. Is it possible that the maximal contraction of the levator when the mouth is opened is compensatory to the weakness of the superior rectus in a fashion similar to Hering's law of equal stimulation of yoke (related) muscles? But this is not completely tenable because not all patients with ptosis associated with superior rectus weakness exhibit the Marcus Gunn phenomenon. Vice versa, not all Marcus Gunn patients present paresis of the superior rectus in the ptotic eye. It would be most enlightening if electromyography could have been done before and after surgery in these five patients to determine the validity of the foregoing statements.

Trauma resulting in desultory nerve repair may explain the acquired form of Marcus Gunn phenomenon, but how about the congenital form? Because of the association of ptosis (despite a normal levator muscle) with weakness of the superior rectus, one would suspect that something is embryologically wrong with the innervation of both muscles. This contention is strengthened further by Falls'[24] report of some inherited cases and by Alexander's[25] report of ptosis and jaw-winking associated with facial asymmetry. Moreover, as already mentioned, the incidence of hypotropia in the eye showing the Marcus Gunn phenomenon is high in this series.

There are other questions regarding the Marcus Gunn phenomenon that demand an answer: Why is it usually unilateral? Why is the left eye more prone to develop it than the right eye? Is there any difference in the etiology between the congenital and the acquired forms? These, including questions on surgical management, remain to be answered more adequately.

I should like to emphasize the following points:

1. It is not wise to consider the ptosis only and to do just a shortening procedure of the levator because, by so doing, the jaw-winking reflex is not broken. The only effective way of interrupting the mandibulopalpebral synkinesia is by resecting a good part of the levator and then utilizing either the frontalis or the superior rectus to elevate the lid.

2. If this is done, it is advisable to do the procedure in two stages: that is, tenectomy of the levator is first performed and, one or two months later, the complete ptosis is corrected. One can then calculate more accurately how much ptosis correction is to be attempted and also can evaluate how completely the mandibulopalpebral synkinesia has been disrupted. Where there is no amblyopia, however, one has to encourage the patient to use the involved eye by purposely raising the ptotic lid with a finger or by patching the good eye.

3. Because of the fact that utilization of either the frontalis or the superior rectus in the elevation of the lid is not as smooth and as efficient as that of the levator, one should refrain from doing surgery in a Marcus Gunn patient unless the jaw-winking is embarrassingly conspicuous.

4. When the ptosis is quite marked but the jaw-winking is minimal, one is confronted with the problem of choosing between two evils. A marked ptosis, of course, poses a greater problem than a minimal jaw-winking that might be worsened only slightly by levator shortening.

5. One should not overlook the heterotropia and/or amblyopia frequently associated with the phenomenon of jaw-winking. Early institution of patching, formal orthoptics, and surgery usually lead to single binocular vision or at least a good cosmetic result.

Summary

The surgical management and results obtained in five cases showing the Marcus Gunn phenomenon are briefly presented. The various concepts regarding its etiology are reviewed. Five points in its surgical management are emphasized, most important of which is that surgery should be avoided in mild Marcus Gunn cases.

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Acknowledgment

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PROBLEMS OF CORTICOGENIC VISION*

A FALSE HOPE FOR THE BLIND

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The title of this paper refers to visual experiences, the stimuli of which lie directly in the cerebrum. In a sense, this is true of all vision. It is a cortical event to which we respond, in the final analysis, not to light. But we mean here only those cortical events over which we have direct control. This means that the stimulus must be electric, or biochemical; mechanical probes are still indirect, in that an electrical event must proceed from them. The control may be through temporary insertions, as during operations, or through permanent implantations.

The visual experience which may sometimes, but not always, result from such stimulation† is known to differ markedly depending upon the region of the brain which is stimulated. In particular, electric stimulation of the temporal cortex of the nondominant side of the body often results in complex hallucinatory experiences, including visual, auditory, verbal, and other sensory components (for example, Penfield and Rasmussen).

Such hallucinations seem to be experiences which the patient has lived through in the past: a party, a dinner, a quarrel, a walk in the country, a song, and so forth. They have all the reality, the color, and the tone of the original event, though the patient (typically under local anesthesia) is generally aware that they are hallucinations, and may even comment on this fact while talking to the doctor. (Many of us can recall dreams which we had while knowing that they were dreams.) However, psychologic complications often arise ancillary to the hallucinations. Strangeness and oddity may invade the experience; the patient may exhibit fear, or marked body image disturbances and paragnosias.

Experiences of such a vivid nature do not occur in patients lacking that heightened sensitivity which is most often concurrent with epilepsy. In particular, such events could not be brought about in a typical blind or near-blind patient. In any event, the hallucinations constitute only a distorted recollection of the past, they bear no relation whatsoever to the present world. That is, the particular form and pattern of the electric stimulation is not reflected in any way in the resultant hallucination. No matter how closely the electric charge may mirror the surroundings of the blind patient, he would experience only his memories. The sole aspect which would be transformed into the experience of the patient is the temporal one: an object enters his vicinity, electric impulses begin, hallucinations begin; the object recedes, electric impulses cease, and the hallucinations stop.

Thus, the idea that electric stimulation of the temporal cortex of a blind person may someday help him to "see" cannot be seriously maintained. It may help him to hallucinate, to recall vividly what he may have seen in the past, but this is all.

Stimulation in other areas of the brain will have different results. All of us, for example, will "see stars", when the back of our head (near the occipital cortex) is jarred with sufficient vigor.

It has also been known for a great many years that the human cortex responds to electric stimulation (Fritsch and Hitzig). This response is quite normal, appearing in all in-

* This manuscript was completed at the American Optical Company Research Center, Southbridge, Massachusetts, where the author was research psychologist. The author is presently on leave to the Technical Optics Section, Imperial College, from the Department of Ophthalmology, University of Miami College of Medicine, Miami, Florida.

† Epileptic patients, for example, have a particularly heightened sensitivity to electric discharges.

dividuals, and is not restricted to those of us with epileptogenic cortical organization. One of the first clinical studies of the direct electric stimulation of the occipital cortex, Brodmann's area 17, by Loewenstein and Borchardt, dates as early as 1918.

The resultant visual experience may be called a cortical "phosphene" or a "photopsia". It is often composed of vague, diffuse, undifferentiated flashes, clouds and fogs of light. Penfield and Rasmussen describe them as follows: ". . . flickering lights, dancing lights, colors, bright lights, stars, wheels, blue, green, and red colored discs, fawn and blue lights, colored balls whirling radiating gray spots becoming pink and blue, a long white mark, et cetera (p. 143)." Sometimes its shape more clearly resembles geometric figures, angles, curves, lines, columns, and planes.

Most often these forms are in rapid and apparently willful motion drifting to one side or the other. However, these shapes are seldom repeatable; it is impossible even to place the electrode repeatedly in the same location —an error of a few millimeters seems to be unavoidable. Moreover, the phosphenes are difficult to describe accurately in the first place. Any person (with an intact retina) who closes his eyes and presses down firmly on his eyelids will get some notion of the effect—this creates a retinal pressure phosphene, however, and not an electric corticogenic one.

It is possible, for our purposes, to make further divisions of the visual cortex. The usual division is into the striate area (Brodmann-17, of which we have been talking), the parastriate area (Brodmann-18), and the peristriate area (Brodmann-19). Duke-Elder comments that B-17 gives rise to simpler, less complex phenomena—true phosphenes—whereas B-18 and B-19 give rise to actual hallucinations (citing Foerster)—such as a friend waving his hand, or butterflies or birds approaching. Penfield and Rasmussen note that phosphenes originating close to the calcarine fissure are colored more often than those originating from secondary visual zones; so, also, are those originating from the occipital pole, as opposed to those originating farther forward.

There is even some slight evidence that local signature exists in the occipital cortex.* Probably, this is dependent upon the point-to-point correspondence between the cortex and the retina (for example, Spalding). In his review of this work, Duke-Elder (p. 3695) reports that: stimulation of the occipital pole gives rise to corticogenic phosphenes localized straight ahead; stimulation of the anterior regions of the striate cortex gives rise to phosphenes in the periphery of the contralateral visual field, which then appear to drift in toward the center; stimulation of the lower lip of the calcarine fissure gives rise to phosphenes which seem to be moving downward from the upper half of the contralateral visual field; stimulation of the upper lip of the calcarine fissure gives rise to phosphenes in the lower periphery of the contralateral visual field.

Penfield and Rasmussen report a careful study of this point. Figure 1 is reproduced from their work. It is a summary of their findings on several subjects, under repeated stimulation. The results are not as confident of local signature as Duke-Elder's comments might lead us to think. Nevertheless, two general trends are clear: (a) as the distance of the electrode from the pole of the occipital lobe is increased, the probability of producing a contralateral phosphene is increased; and (b) as the end of the calcarine fissure (the occipital pole) is approached, the probability of producing a "nonlateralized" phos-

* Local signature is a term usually applied to the retina. It is held that each cell or tiny collection of cells (Panum's areas) possesses a fixed (specific) quality of direction (sign of location) with respect to the fovea. It is the cell stimulated, and not the physical location of the stimulus, which determines where an object will be seen. In applying this term to the brain, I wish to retain the same meaning, that the cell stimulated would have a particular spatial value associated with it.

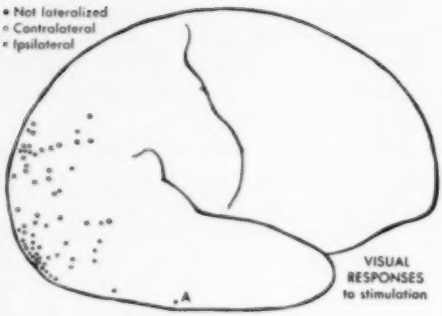

Fig. 1 (Shipley). The local signature of the cortex. (From Penfield and Rasmussen. Reproduced with the kind permission of the authors and the Macmillan Company, publishers.)

phene is increased. Ipsilateral phosphenes were not obtained with sufficient frequency so as to allow hypotheses as to their origin.

In no case is it reported that two, or more, of these phosphenes can be seen simultaneously. Moreover, Lashley's (a) reason for affirming the impossibility of preserving local signature by direct electric stimulation of the optic nerve—namely, the extremely close and complex interweaving of fibers—probably applies to the cortex with even more force. The objection would be insurmountable if the theory of the cortex as a volume-conductor should prove true (Köhler, Held, and O'Connell).

There are, of course, many kinds of blindness. It is grossly misleading to discuss this issue in generalities. If the neurology is sound and only the preretinal media are at fault, as is true in a great many instances, then we would expect the patient to exhibit some sensitivity to retinal phosphenes. And, indeed, Schlodtmann, long ago, has shown this to be true for the pressure phosphenes. More recently, this finding has also been demonstrated with the electric phosphene (known as early as 1775—see LeRoy). A short review of his work may be found in Haskins.

But these are retinal phenomena, the problem of corticogenic phosphenes is more complex, and almost nothing is known about them in blind patients.*

In many cases of blindness, the brain itself is fully intact, while one or a number of the precortical media is at fault. Even in cases of extensive brain-damage rudimentary aspects of vision may remain. Form and color perception may be destroyed, for example, while brightness and motion perception may remain virtually intact (for example Lashley, b). Color discrimination is extremely vulnerable to cortical injuries, whereas gross sensitivity to position—or rather to changing position—is particularly resistant (for example, Stenvers).

Hence, constantly confronted with one or another case of absolute blindness of peripheral origin (for example, enucleation), the hope periodically arises in us that corticogenic vision may be found useful. We have already commented on the obvious inadequacy of the temporal cortex for this purpose. Moreover, as we have also implied, corticogenic phosphenes from the occipital cortex have never been induced in a form which resembles the form of the stimulation other than in its temporal aspects, that is in its off-and-on characteristics.

This is the essence of the problem. To my knowledge, it has not yet been possible to produce even two pure and distinguishable phosphenes simultaneously. It is true that relative spatial positions are recognizable. One can "see" something to the right or to the left of another, but this is within the pattern and flow (one can tell direction of flow) of a single phosphene. While such rudimentary local signature does exist, I cannot find, even in the literature on sighted subjects, whether the simultaneous evocation of two or more discrete phosphenes has ever been successfully achieved. Judging from the informational capacity of our other senses, at least 10 discrete phosphenes (not two) would be

* Search of the scientific literature, for example, has revealed only a few casual references to this subject.

necessary in order to establish a "cortical alphabet" which could be used for most normal tasks.

This alphabet of 10, however, would not allow for the sensation of visual configurations, but only for their labeling—as a square may be one short and two long phosphenes or one phosphene straight ahead and two on the left side. The preservation of actual forms is an entirely different thing. Lashley (c) finds, with animals, that a minimum of 1,200 central visual fibers must be functioning in order that the simplest geometric forms may be distinguished. On the basis of the well-known arborization in man, of about 100:1 between cortex and retina, we might guess that something of the order of 100,000 cortical cells must be functioning in order for us to perceive visual forms. This is, in fact, a very small number. The point is, however, that this may probably be the order of magnitude of the number of electrodes necessary even to approach the simulation of cortical local signature—a clearly impossible number.

There is some slight indication in the literature that intensity changes may be sensed —but even this is not yet too clear. I do not think that frequency effects have been studied. Probably, it is not too much to hope that with frequency, intensity, number, and place, we might be able to elaborate an alphabet of say 10 labels—more than this is idle thought.

In any case, it will at best be a great many years before the profession will be able to give any sort of corticogenic vision to a blind person by the permanent insertion of electrodes into his brain. Not only is the successful installation of permanent electrodes a new art (Delgado), but, more important, it is a highly speculative hope that we could ever create useful "vision" by this means.

Recently, however, some work in this area was reported by Dr. John C. Button. (Conference on "Electrical techniques in biology and medicine," Boston, Mass., Nov. 7, 1957; see also *New York Times*, Nov. 21, 1957; I have not yet seen it reported in the scientific literature.) Unfortunately, the publicity surrounding this work carried a great number of glaring misconceptions. The fact that report of this work has since appeared in many other places (electronics magazines, optometric literature, and particularly in the magazine *Listen*, which is circulated to the blind in a braille edition), necessitates a direct comment.

In this experiment several electrodes were implanted in the occipital cortex of a woman, totally blind for 18 years. The cause of the blindness seems to have been a tumor which brought about a complete neurologic blockage somewhere below the cortex. Apparently, this woman responded "normally" to electric current, and described a "typical" corticogenic phosphene.

The importance of the work is just and only this fact: a phosphene was induced which, at least superficially, resembled that induced in sighted subjects. Hence, whatever structure(s) it is in the brain which engenders the phosphene, this was not destroyed by 18 years of blindness. It cannot be claimed that this structure functions normally, only that it has not entirely ceased its function.

In view of the difficulty which sighted subjects have in describing phosphenes, it is not at all certain that those which this patient experienced actually did resemble those experienced by normal subjects. The implication that the cortex was completely sound is specious.

Animals reared in total darkness clearly exhibit peripheral and central degeneration (Riesen; Clark); signs of cortical disturbances are common in many kinds of blindness (Parsons-Smith), and are used for diagnostic purposes (Case). Therefore, in most cases of long-term blindness it is probably best to assume some cortical degeneration. Lashley (c) concludes, for example:

> Where the retina has been destroyed as in a great number of cases of blindness which cannot be treated surgically, the optic nerve undergoes degen-

eration and is inexcitable. The second-order cells of the lateral geniculate nucleus also undergo a partial degeneration, losing their Nissl granules and showing some shrinkage. Ultimately the granular cells of the visual cortex also show regressive changes. The nature of these changes . . . and the degree of functional loss which they entail are not understood, but it is probable that the receptive layer of the visual cortex . . . loses its capacity for normal functioning (p. 503).

Each type of blindness must be examined independently. In many cases of total blindness, photopsiae (Duke-Elder) and visual hallucinations (Lende gives a whole bibliography on these, pp. 143-146) are commonly reported; indeed, more commonly than not. In fact, these often cause greater distress than the actual loss of sight. Severing the optic nerve, as in enucleation, almost invariably give rise to flashes of light", regardless of the number of years that the patient has been blind. Many long-term blind patients will experience visual sensations when struck on the back of the head. In all these cases, it seems to me, we cannot escape the conclusion that some cortical functioning has remained. It is not clear, therefore, just what physiologic significance, if any at all, can be credited to Button's findings.

Moreover, in cases of relative blindness (cases in which some light sensitivity has remained), reasonably good vision has been achieved by these patients even after 50 years, for example through belated cataract operations. Such cases have been in medical records for many hundreds of years. (See Lende for a helpful bibliography on this: "Sight after blindness," pp. 123-128.) Hence Button's finding that the cortex is particularly resistant to degenerative changes, though certainly well taken, is not unknown in the field.

Button went further than this, however, and wired a photocell to the electrodes. The patient held this photocell in her hand and used it as a wand. Only when she turned it toward the light did she experience the corticogenic phosphene. In this way the completely false implication was made that the patient actually "saw" the light. We all must sympathize with the patient's enthusiasm, but she surely did not see the real light, there in the room; she only perceived the effect of an electric disturbance in her brain. In principle, light had nothing whatsoever to do with what the patient experienced. This, as we have said, is the nature of corticogenic vision. If, instead of a photocell, a thermocouple had been put on the end of the wires, the patient would have "seen" the heat of the radiator; if a microphone pickup had been used, she would have "seen" the words which were spoken. And, as far as knowing where the light was, a photocell which rang a bell when pointed at the light, or which tapped the patient on the finger, would have transmitted exactly the same amount of information. The brain need not have been invaded at all.

Moreover, it has not been shown that two patterns could be obtained simultaneously, and the difference between one vague impression of luminance and a differential response to two lights (that is, to the form of the stimulus) is an infinity. As we have shown, it is only this latter finding which would hold hope for a visual aid.

In summary then, our knowledge of induced corticogenic vision is in a very primitive state, its use as an aid to the blind is only a dim hope at best. Work in this area should be undertaken with great restraint and extreme scientific caution.

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CONGENITAL ECTROPION AND DISTICHIASIS*

Etiologic and hereditary factors: A report of cases and review of the literature

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Congenital ectropion and distichiasis, more especially the former, are anomalies of the eyelids to which the much abused term "rare" can properly be applied.

Ectropion

Most textbooks either do not mention the congenital variety of ectropion at all or devote little space to it. Duke-Elder,[1] for instance, describes secondary eversion of the lid margins as a congenital phenomenon associated with microphthalmos and buphthalmos and secondary eversion of the lower lids associated with orbitopalpebral cysts but notes that primary ectropion is very rare indeed. Sorsby[2] mentions the possible existence of congenital entropion in his detailed and extensive presentation of congenital anomalies of the eyes and adnexa. Mann,[3] after noting that both entropion and ectropion are exceedingly rare as congential defects, says of ectropion that it involves the upper eyelids more often than the lower and may be associated with either an abnormality or complete absence of the tarsal plate. If the lower lid is involved, she continues, microphthalmia is usually present, associated with an orbital cyst. If the upper lid is involved, there may be a deformity of the skull; her reason for the last statement, which is not borne out by the recorded cases, is not apparent.

An exhaustive review of the literature (including, as a precaution, the literature of congenital entropion) has revealed only 12 recorded cases of congenital ectropion. These

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few reports do not make clear the anatomic factors involved in the production of this anomaly, nor do they pay particular attention to the role of the gene as an etiologic factor. There are, in fact, few clear-cut statements in these papers about the direct causation of congenital ectropion, some writers apparently failing to realize that it is the expression of a variety of causes rather than a single pathologic entity.

To clarify this confused situation, the following etiologic classification is suggested:

1. Congenital ectropion due to absence or anomaly of the tarsus.

2. Congenital ectropion (congenital eversion of the eyelids, the nomenclature suggested by Ostriker and Lasky[4]) due to eversion of the eyelids during passage through the birth canal.

3. Congenital ectropion due to changes in the skin of the lids, such as would be associated with hyperkeratosis or ichthyosis congenita.

4. Congenital ectropion due to microphthalmia and orbital cysts.

Groups 1 and 2 represent true primary congenital ectropion. Groups 3 and 4, in which both causative factors and diagnosis are usually clearcut, represent the secondary congenital variety.

Distichiasis

The literature on congenital distichiasis, although it is considerably more extensive than the literature on congenital ectropion, is still scanty, and in only a few reports is it mentioned that congenital ectropion and congenital distichiasis may coexist.

After encountering a case of combined congenital ectropion and distichiasis, I found myself fortunate enough to be able to study three generations of the patient's family, including her own; most of the members could be examined personally. Ten of the patient's relatives suffered from congenital ectropion and nine from distichiasis, either as an isolated entity, or, in seven cases, in combination with ectropion.

This remarkable concentration of cases in a single family provided an opportunity, apparently not previously available, to study the probable mode of inheritance of congenital ectropion and congenital distichiasis. The microscopic studies carried out in two cases also made it possible to implicate a defect of the tarsus as a possible causative agent in congenital ectropion.

Case studies

This investigation, which took place in Puerto Rico, was stimulated by the observation of an 18-year-old girl (hereafter called the propositus) who showed marked ectropion of both lower eyelids associated with distichiasis of the upper eyelids. When questioning revealed that many of her ancestors and living relatives apparently presented similar anomalies, a detailed study was undertaken of all members of the family.

The investigation (fig. 1) revealed 19 relatives of the propositus, in four generations, four of whom were dead. Thirteen of the 15 living relatives were personally examined, and information about the other two was obtained from Dr. Edmund B. Spaeth, who had operated on them.

Although there was no history of consanguinity in the family, the spouses of the married members were included in the investigation. As examination revealed no evidence of ectropion or distichiasis in any of them, they are omitted from the record except for the (deceased) grandfather of the propositus (generation I, 1), who is included to complete the chart. It was reported that his eyes were normal.

The (deceased) grandmother of the propositus (I, 2) was said to have had the same

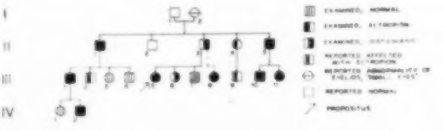

Fig. 1 (Picó). Occurrence of congenital ectropion and distichiasis in family of 20 members.

types of "small" eyes seen in several living members of the family. In the next generation, a boy (II, 2), who had died at the age of seven years, apparently had normal eyes. A female in this generation (II, 4), who had died at the age of 35 years, had apparently had an extreme ectropion of all four eyelids since birth; no information was available about a possible distichiasis in this case.

The routine examination in the 14 members of this family examined personally consisted of external examination and ophthalmoscopy, usually accompanied by refraction. Special studies consisted of (1) inspection of the position and appearance of the eyelids; (2) palpation of the lids to determine the condition of the tarsus; (3) investigation of the margin of the eyelids, including slitlamp examination, with a special search for aberrant eyelashes and the openings of the meibomian glands; (4) determination of the length of the palpebral fissure; (5) determination of the height and vertical extension of the upper eyelids by Fuchs' technique; (6) a study of the conjunctiva; (7) a study of the cornea for opacities; (8) tonometry.

Case histories

The case histories which follow, as well as the case reports from the literature, include only the findings important in relation to ectropion and distichiasis. Unimportant and irrelevant negative findings are not included.

Case 1 (III, 5; fig. 2).

This 18-year-old white girl, the propositus, was first seen December 31, 1952, with a history of ectropion of the lower eyelids since birth. She complained of photophobia and irritation of both eyes. On questioning, she stated that several other members of her family suffered from the same condition.

Examination showed complete ectropion of both lower eyelids, easily corrected by pulling the temporal third of the lids up and out. The skin was loose and flaccid but otherwise normal in appearance. There was no evident shrinkage of the subcutaneous tissues, but on palpation the tarsus could not be demonstrated and the lids seemed thinner than normal. Vertical extension of the lower lids

Fig. 2 (Picó). Ectropion of lower eyelids before operation (III, 5, Case 1).

from the lower border of the orbit to the border of the replaced lids was apparently normal.

Function of the orbicularis muscle in both lids on both sides seemed normal, but when the girl was told to close her eyes, as if she were asleep, closure was not complete and the lowermost portion of the cornea and a portion of the eyeball were left exposed.

The lower eyelashes were in normal position at the anterior border of the lid margin, but they were much smaller, thinner, and paler, as well as less numerous, than the upper eyelashes. The openings of the meibomian glands could not be seen in either upper or lower lid, and no lashes were found at the posterior border of the margin of the lower lids, where the meibomian orifices are normally present.

The upper eyelids were normal in position and apparently normal tarsi could be demonstrated. The anterior row of eyelashes was normal, but on each upper lid, just above the outer canthus and very close to the posterior border of the lid margin, was a bunch of eight or 10 apparently normal cilia. Close to the lacrimal punctum on each upper lid, also near the posterior border of the lid margin, were three rather small eyelashes.

The palpebral fissures were narrow horizontally. The following measurements of the upper lids were obtained by Fuchs'[8] technique: right upper eyelid: length 18 mm., vertical extension 25 mm., coefficient, 1.39; left upper eyelid: length 20 mm., vertical extension 27 mm., coefficient 1.35.

The palpebral conjunctiva of both lower lids was congested. Some ciliary injection was present in both eyes. Corneal nebulas were present in the lower quarter of the right eye and the lower third of the left eye; a few small, superficial corneal ulcers were seen in these areas.

Chloretetracycline (Aureomycin) ointment was used until operation, which was performed on the left eye April 17, 1953, by the Kuhnt-Szymanowski technique, as follows:

The entire margin of the lower lid was split just posterior to the line of lashes. Since absence of the normal firm structure of the tarsus made it impossible to remove a triangular wedge, apex downward, as the Kuhnt technique requires, the procedure was altered, and a wedge of tissue was removed from the middle of the lid, including the conjunctiva, connective tissue, orbicularis muscle, and skin. Then a triangular portion of skin, with the base directed upward and outward, was re-

Fig. 3 (Picó). Appearance of patient shown in Figure 2 after surgical correction of ectropion.

moved at the outer canthus, according to the Szymanowski technique. The triangular wound was closed with three interrupted sutures of 6-0 silk in the conjunctival aspect, with a single additional suture at the lid margin. The anterior flap of the split lower lid was undermined, moved temporally, and sutured to the upper and temporal aspects of the triangle with interrupted 4-0 and 6-0 silk sutures. Before the sutures were tied, the lashes now in the upper border of the triangular area were removed and this area was scraped.

At the conclusion of the operation the ectropion seemed, if anything, to be overcorrected. Twelve days later, however, after the sutures and bandages were removed, it began to reappear, though in considerably less pronounced form. The scar in the outer canthus gradually widened, eventually assuming the appearance of a flat keloid. The disappointing result was thought to be due to the absence of the tarsus.

On July 16, 1954, the procedure just described was repeated on the left eye and was also performed on the right eye, in which the tarsus, as in the left eye, was missing. The final result in the left eye was excellent. In the right eye, the result was poor, but the ectropion was completely corrected after the operation was repeated on July 22, 1955.

The patient is well satisfied with the improvement in her appearance (fig. 3). Photophobia is no longer troublesome and the keratitis has disappeared, but corneal nebulas are still present.

The pathologist reported as follows on the tissue removed at the first operation on the left eye:

This wedge-shaped piece of tissue fixed in formalin measures 1.1 by 0.6 by 0.4 cm. One of its broad surfaces is covered by a brown, smooth membrane, which could represent skin and which continues over an acute border and upward on the opostie side, where it becomes white and resembles a mucosal lining. When the specimen is sectioned perpendicularly to these two surfaces, grayish-white tissue without distinct layering is exposed.

Microscopic examination shows that the palpebral conjunctiva, which has undergone slight broadening, contains practically no mucus-secreting cells and shows slight hyperkeratosis (fig. 4). Beneath it is connective tissue, which is slightly hypervascularized. No tarsal plate is evident, and there is a total absence of meibomian glands. The connective tissue shows elastic fibers and well-developed collagen (fig. 5).

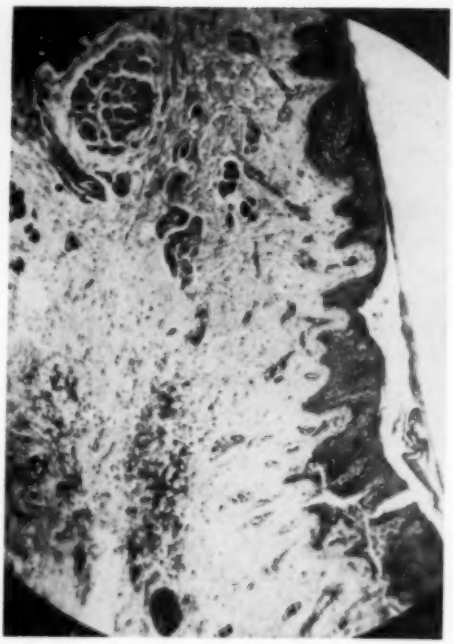

Fig. 4 (Picó). Photomicrograph (Case 1) showing broadening of conjunctiva with slight hyperkeratosis and absence of tarsal plates and meibomian glands. The muscle of Riolan is included in the specimen. The hemorrhage into the connective tissue occurred at operation. (×100).

CASE 2 (II, 3; fig. 6).

This 47-year-old male, father of the propositus, was first seen on March 2, 1954, with the chief complaint of inflammation of both eyes due to inturned eyelashes. The condition had been present since childhood, and periodic epilation was carried out by a relative. The patient's vision had been poor in early childhood, and he had been operated on for congenital cataracts at the age of 10 years.

Examination revealed the eyelids normal in position, with no ectropion or entropion, and of normal thickness. Palpation revealed the tarsus to be present. Although many lashes were missing in the upper lids, each lid had the normal anterior row of lashes. The usual openings of the meibomian glands were not demonstrable in either the upper or the lower lids. The posterior border of the margin of the right lower eyelid was ill defined and rounded; close to this border 10 rather thin and short eyelashes were present in the nasal half and eight in the temporal half. These lashes lay chiefly along the margin of the lid, but some drooped forward and their shafts intermingled with the shafts of the anterior rows of lashes. Similar findings were demonstrable in the left lower lid, on which only 12 abnormal lashes were observed.

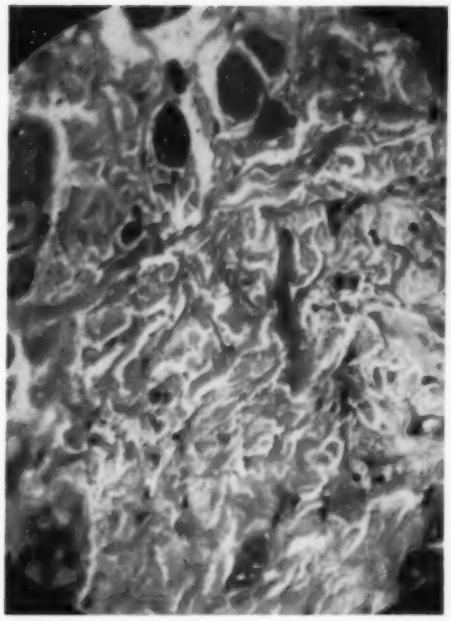

Fig. 5 (Picó). Photomicrograph (Case 1) showing replacement of tarsal plate by connective tissue. Note fibers of muscle of Riolan (×460).

The 12 to 15 additional lashes present in each of the upper lids arose close to the inner border of the lid margin, in the normal location of the meibomian orifices. Eight to 10 of these aberrant lashes were in the temporal half of the lid and four to six in the nasal half. Some of them were of the lanugo type and were so pale, thin, and small that they could be seen only with the loupe or the slitlamp. Most of them were turned inward and rubbed against the eyeball, explaining the discomfort and irritation of the bulbar conjunctiva of which the patient complained.

The palpebral fissure was unusually narrow horizontally, measuring only 20 mm.; the finding suggested blepharophimosis. Measurements of the upper eyelids by Fuchs' technique were as follows: right lid: height, 17 mm., vertical extension 25 mm., coefficient 1.47; left lid: height, 19 mm., vertical extension, 27 mm., coefficient 1.44. Although the coefficient was below the normal level in both upper eyelids, the lids could be completely closed and no lagophthalmos was present.

The cornea was clear, but examination with the slitlamp showed some vascularization for two to three mm. next to the limbus.

The patient was aphakic, and only small remnants of the lens capsule could be seen in each eye behind the iris.

Electrolytic epilation of the eyelashes arising from the posterior border of the margin of the upper eyelids was done with some difficulty; even with magnification, some of the lashes were difficult to demonstrate. Three epilations were necessary to eliminate them all and to relieve the discomfort and irritation of the bulbar conjunctiva.

CASE 3 (III, 6; fig. 7).

The 23-year-old sister of the propositus, who was examined on February 10, 1956, denied all complaints referable to her eyes except that they tired easily. On further questioning, she stated that her lashes had turned in since childhood.

Examination showed all lids normal in position. Palpation revealed normal tarsal plates in the upper lids and normal openings of the meibomian glands close to the posterior border of the lid margins. In the lower lids, which were abnormally thin, the tarsal plates were either absent or extremely deficient in thickness, and no openings of the meibomian glands could be demonstrated.

On each of the upper lids there was a normal row of eyelashes along the anterior border of the margin. On each of the lower lids there was also a normal row of lashes, though they were rather short and pale. Near the posterior border of each lower lid was a row of 15 aberrant lashes, all of them short, whitish, and thin. In the right lid, these lashes occupied only the nasal two thirds; in the left lid they extended almost to the outer canthus. All aberrant lashes were inclined along the border of the lid, except for three on each side in the region of the lacrimal punctum; here they turned inward

Fig. 6 (Picó). Absence of ectropion in patient with distichiasis, which is not noticeable in this photograph (II, 3, Case 2).

Fig. 7 (Picó). Absence of ectropion in patient with distichiasis, which is not noticeable in this photograph (III, 6, Case 3).

Fig. 8 (Picó). Normal eyelids (III, 7, Case 4).

and brushed against the bulbar conjunctiva, though not enough to produce inflammation.

CASE 4 (III, 7; fig. 8).

The 15-year-old brother of the propositus was examined in July, 1956. He had been committed to a government institution for children, in which he was classified as a moron.

Examination showed the eyes to be normal in all respects. The eyelids were also normal in position, thickness, structure, tarsal plates, eyelashes, and openings of the meibomian glands.

The palpebral fissures measured 28 mm., horizontally and 10 mm. vertically. Measurements of the upper eyelids by Fuchs' technique were the same on both sides: height, 22 mm.; vertical extension, 28 mm. The coefficient, 1.3, was below the normal of 1.5 set by Fuchs, but this boy did not have lagophthalmos and both lids were completely closed when he was asleep.

CASE 5 (III, fig. 9).

This 14-year-old girl, an illegitimate daughter of the father of the propositus, was examined on April 2, 1956. Although her only complaint was photophobia, her mother stated that her eyelids were not completely closed when she was asleep.

Examination revealed slight ectropion of both lower lids, with a possible bilateral weakness of the orbicularis muscle. The lower lids were abnormally thin, and no tarsal plates could be felt on palpation.

The normal openings of the meibomian glands could not be demonstrated in either upper or lower eyelids. No aberrant lashes were present in the lower lids. The upper eyelids were normal in position and thickness, and tarsal plates were apparently normal. On each upper lid there was the usual row of lashes, but a second row, of about 15 lashes, was present in the temporal third. These aberrant lashes, which were smaller, paler, and thinner than those of the anterior row, arose from the posterior border of the lid margins where the openings of the meibomian glands are usually situated; no openings were seen elsewhere along the margin of the upper lids. The direction of the aberrant lashes was the same as that of the anterior rows of lashes, and they did not touch the eyeball when the lids were opened or closed. The eyelids did not close completely when the child was told to pretend that she was going to sleep.

Fig. 9 (Picó). Slight ectropion of lower eyelids. Distichiasis is present in the upper lids but is not noticeable in this photograph (III, 8, Case 5).

The palpebral fissures were narrow, measuring 22 mm. horizontally on the right and 23.5 mm. on the left. The vertical measurement was eight mm. on each side. Measurements of the upper lids by Fuchs' technique were the same on each side: height, 18 mm.; vertical extension, 24 mm.; coefficient, 1.33. The coefficient was below the normal of 1.5.

There was some congestion of the bulbar conjunctiva in each eye and slight ciliary injection inferiorly on the right side. Some opacities were present in the lower third of each cornea. Photophobia was extreme.

The conjunctivitis in both eyes, the keratitis on the right side, and the corneal opacities on the left side were considered due to the slight ectropion rather than to the distichiasis. Therefore, after treatment of the active keratitis in the right eye with an ointment containing tetracycline and hydrocortisone, a Kuhnt-Szymanowski operation was performed on the right lower lid on June 1, 1956. As in Case 1, no tarsal plate was found. The wedge of tissue removed from the middle of the lid included all tissues from the palpebral conjunctiva up to the skin in that area; the skin was not removed, as it had been in Case 1.

The pathologist reported as follows on the specimen:

This wedge-shaped piece of tissue, fixed in formalin, measures 0.6 by 0.5 by 0.3 cm. One side, which is covered by conjunctiva, is white, smooth, and glistening. The opposite side, which is the apex of the wedge, is brown and slightly uneven. On section, there is no layering evident, and the predominant coloration is gray.

On microscopic examination, the epithelial layer of the conjunctiva is seen to be very slightly broadened. It contains a few mucus-secreting cells. A slight infiltration with plasma cells, lymphocytes, and a few large mononuclear cells is observed immediately beneath the epithelium. The subconjunctival layer of fibrous tissue is broader than in the other case studied histologically (Case 1) and is likewise totally devoid of meibomian glands (fig. 10). The subconjunctival connective tissue is denser in some areas than in the other specimen; it contains numerous broad collagen bundles, as well as elastic fibers, but beneath the conjunctiva the tissue is loose and edematous (fig. 10). The root of a hair follicle, implanted in the deepest part of the

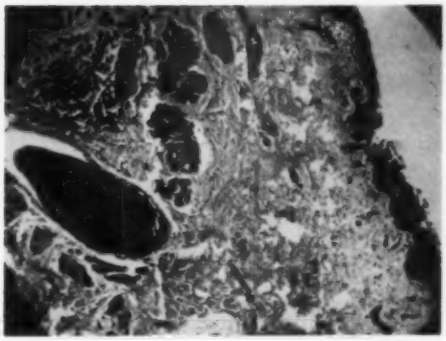

Fig. 10 (Picó). Photomicrograph (Case 5) showing absence of meibomian glands and replacement of tarsal structures by lax fibrous tissue (×100).

orbicularis muscle, can be seen near the lid margin.

Results of surgery were good in this case. The right lower lid was better approximated to the eyeball, and the right eye was well closed when the child was asleep. The same procedure will shortly be carried out on the left side.

CASE 6 (II, 1; fig. 11).

The 56-year-old lawyer, paternal uncle of the propositus, complained of burning of the eyes and photophobia when he was examined on August 2, 1956.

Both upper eyelids were normal in position, but a definite ectropion was present in both lower lids. All four lids were much thinner than normal. No tarsal plates could be demonstrated in the lower lids, and the plates palpated in both upper lids were thin and small. Even with the slitlamp it was not possible to demonstrate openings of the meibomian glands in either upper or lower lids. A group of four or five lashes arose from the posterior border of the margin of each upper lid, about one to two mm. above the outer canthus, and similar groups were present two mm. further superiorly. These aberrant lashes, which were similar in appearance to those in the normal anterior rows in the upper lids, were inturned and rubbed against the eyeballs.

Slitlamp examination revealed about 15 aberrant lashes in the lower lids, arising from the margin close to the posterior border, along which they were distributed. They were not inturned and did not brush against the eyeball when the lids were closed.

The lids were apparently closed normally when the patient was asleep.

The palpebral fissures measured 24 mm. horizontally and seven mm. vertically. In this patient, as in several other members of the same family, the small fissures, especially the short horizontal measurements, gave the impression of small eyes. Measurements of the upper lids by Fuchs' technique showed the height of each lid to be 17 mm. and the vertical extension 26 mm. The coefficient, 1.53, was low for the patient's age.

There was some congestion of the bulbar conjunctiva in both eyes, and a small corneal nebula was present in the left eye, next to the lower limbus.

Electrolysis of the inturned eyelashes has been recommended but has not yet been accepted.

CASE 7 (III, 1; fig. 12).

This 26-year-old dental student, a cousin of the propositus, was examined on August 2, 1956. His chief complaint was inturned eyelashes, with irritation of the eyeballs. Electrolysis of some of the lashes in the nasal third of both upper eyelids about a year earlier had not been successful; most of them had soon reappeared.

Fig. 11 (Picó). Slight ectropion of lower eyelids. Note that small palpebral fissures give impression of "small" eyes. Distichiasis was present in both upper and lower lids but is not noticeable in this photograph (II, 1, Case 6).

Fig. 12 (Picó). Slight ectropion of lower eyelids. Distichiasis was present in all four lids but is noticeable only in the nasal third of each upper lid (III, 1, Case 7).

Examination showed very slight ectropion in the temporal half of each lower lid. All four lids seemed much thinner than normal. In the upper lids, undeveloped tarsal plates could be palpated about four mm. upward from the border, but no definite tarsal structures could be demonstrated in either lower lid.

All four lids presented the normal anterior row of lashes. In the nasal half of each upper lid were 15 aberrant lashes, which arose from the posterior border of the lid margin. They were somewhat thinner and shorter than the normally placed lashes

Fig. 13 (Picó). Slight ectropion of right lower eyelid. Note distichiasis present in upper lid (III, 1, Case 7).

and the shafts of most of them rubbed against the eyeball. There were three additional aberrant lashes in the right upper lid, close to the outer canthus (fig. 13), and eight similarly placed aberrant lashes in the left upper lid. In the nasal half of each lower eyelid 10 or 12 small, white, lanugolike hairs arose from the posterior border of the lid margin; they were inclined along the lid margin and therefore did not rub against the eyeball. The normal openings of the meibomian glands could not be demonstrated in any of the eyelids. The patient closed his lids well while he was asleep.

Measurements of the upper lids according to Fuchs' technique showed the height of each lid to be 22 mm. and the vertical extension to be 30 mm. The coefficient was 1.36.

The bulbar conjunctiva was slightly injected. There were no corneal opacities.

Electrolytic epilation of the aberrant inturned lashes in the upper lids was done on August 7, 1956, with excellent results.

Case 8 (IV, 1; fig. 14).

This four-year-old second cousin of the propositus was examined on August 2, 1956. She had no complaints referable to her eyes, and all findings (lids, lashes, openings of the meibomian glands) were entirely normal. Measurements of the upper lids by Fuchs technique showed the height to be 18 mm. and the vertical extension 26 mm. The coefficient, 1.45, was slightly below the established normal.

Case 9 (IV, 2; fig. 15).

This two-year-old boy, a second cousin of the

Fig. 14 (Picó). Normal eyelids (IV, 1, Case 8).

Fig. 15 (Picó). Ectropion of some degree in all four eyelids. The distichiasis present in the lower lids is not seen because the aberrant lashes are pale and thin (IV, 2, Case 9).

propositus, was examined on August 2, 1956. According to his parents, ectropion had been present in all four eyelids since birth, and the eyelids did not close completely when he was asleep.

Examination showed marked ectropion in all four lids, most evident and most complete in the lower lids, in which the palpebral conjunctiva was completely exposed. In the upper lids, the anomaly was only partial when the eyes were open but became complete when they were closed (fig. 16). Tarsal plates could not be demonstrated in any of the four lids.

On the upper lids the growth of eyelashes was entirely normal. On the lower lids the normal, anteriorly placed lashes were thin and pale, and on each side some 15 or 20 lanugolike hairs arose from the posterior border of the margin. Some of these aberrant lashes were turned inward and rubbed against the eyeball. Normal openings of the meibomian glands were not present in any of the lids.

The palpebral fissures measured 22 mm. horizontally and eight mm. vertically. The upper eyelids were definitely shorter than normal. By Fuchs' technique the height was 17 mm. and the vertical extension 20 mm., which gave a coefficient of 1.12.

The palpebral and bulbar conjunctivas were congested. Some opacities were present in the lower quarter of each cornea.

Plastic surgery has been recommended to this patient. The abnormal shortness of the eyelids will require tarsorrhaphy, with skin grafts applied to all four lids.

Fig. 16 (Picó). Ectropion of upper eyelids shown in Figure 15, more complete when lids are closed.

Fig. 17 (Picó). Normal eyelids (III, 3, Case 11).

Fig. 18 (Picó). Slight ectropion of lower eyelids. Distichiasis was present in all four eyelids but in this photograph is seen only in the upper lids close to the outer canthus (II, 5, Case 14).

CASE 10 (III, 2).

This 24-year-old cousin of the propositus was not available for personal examination but Dr. Edmund B. Spaeth has informed me that he operated on him, when he was 21 years of age, for congenital ectropion of all four lids, which had been evident since birth. He had a superficial punctate keratitis as the result of incomplete closure of the lids and exposure of the cornea. A severe chronic photophobia made it impossible to secure photographs. The ectropion was corrected with skin grafts, without resection of any tissue, with excellent results. No information is available in this case concerning the condition of the tarsus or the presence or absence of distichiasis.

CASE 11 (III, 3; fig. 17).

The 10-year-old female cousin of the propositus was examined in May, 1956. No abnormalities of any kind were evident. Measurements of the height and vertical extension of the upper eyelids were not secured.

CASE 12 (III, 4).

The seven-year-old male cousin of the propositus was examined in May, 1956. All findings were normal. Measurements of the height and vertical extension of the upper eyelids were omitted. Because of the child's lack of co-operation, photographs could not be secured.

CASE 13 (III, 9).

The 16-year-old cousin of the propositus was not available for personal examination, and information concerning him was obtained from Dr. Edmund B. Spaeth, who operated on him, at the age of 15 years, for ectropion which had been present since birth and which affected all four lids. The boy had a superficial punctuate keratitis, due to incomplete lid closure, and a chronic severe photophobia, which made it impossible to obtain photographs. The ectropion was corrected with skin grafts, without resection of any tissue, with excellent results. No information is available concerning the condition of the tarsal plates or the presence or absence of distichiasis.

CASE 14 (II, 5; fig. 18).

This 44-year-old lawyer, uncle of the propositus, was examined in May, 1956. He complained of some irritation in the right eye and considerable photophobia. He had had slight ectropion of both lower lids since birth, and had a history of inflammation in the left eye in infancy and again in 1938 and 1946.

Examination revealed slight ectropion of both lower lids, more pronounced in upper gaze. No tarsi were demonstrable in the lower lids. In the upper lids the tarsal plates were slightly thinner than normal and extended less far upward than normal.

In addition to the usual anterior row of lashes present in the upper lids, a bunch of eight lashes arose from the posterior border of the lid margin, two mm. from the outer canthus. They were inclined toward the eyeball and sometimes rubbed against it (fig. 19). The anterior lashes on the lower lids were whitish and weak-looking and were far fewer than the number usually present. In the middle third of each of the lower lids were some almost invisible, lanugolike lashes which arose from the lid margin, close to the posterior border. The normal openings of the meibomian glands could not be demonstrated in any of the four eyelids.

The palpebral fissures were narrow in the horizontal diameter, measuring only 24 mm. on each side. Vertically, the lids opened 10 mm.

There was some congestion of the bulbar conjunctiva in each eye. In the right eye a faint corneal nebula extended upward from the limbus for 3.5 mm. In the left eye the nebula, which occupied the lower two thirds of the cornea, was denser than on the right and superficial vascularization was present.

Fig. 19 (Picó). Left eye of patient shown in Figure 18, showing distichiasis in upper eyelid near outer canthus. Note dense vascularized corneal macula.

Fig. 20 (Picó). Slight ectropion of lower eyelids. Distichiasis present in all four lids is not noticeable in this photograph (III, 10, Case 15).

CASE 15 (III, 10; fig. 20).

This 14-year-old cousin of the propositus was examined in New York on July 6, 1956, through the courtesy of Dr. Ramón Castroviejo, who also made available to me the previous medical record. The boy had a history of irritation of both eyes in early childhood but had no complaints at the time of the 1956 examination.

A very slight ectropion was present in both lower lids, in neither of which was a tarsal plate demonstrable. In the upper lids there was no evidence of a tarsal structure in either the nasal or temporal third, and only a very small plate in the middle third.

The normal anterior row of eyelashes was present in all four lids. On the right upper lid four aberrant lashes arose from the posterior border of the margin, about one cm. from the outer canthus, and a single lash, of similar origin, was present about five mm. from the inner canthus. There were no aberrant lashes on the right lower lid. On the left upper lid there was an aberrant row of about 40 lashes, similar in color, length, and direction to the normal anterior row. They arose from the posterior border of the lid margin and were localized in the middle three fifths of the border. On the left lower lid a single pale, thin lash arose from the posterior border of the lid margin, in the middle third. None of the aberrant lashes brushed against the eyeball. Normal openings of the meibomian glands were not observed in any of the four lids.

The margin of the left upper eyelid was normally placed except when the patient looked upward; then a slight entropion was evident, but even under these circumstances, the extra row of eyelashes, though they were in close approximation to it, did not rub against the eyeball.

The palpebral fissures were narrow in the horizontal diameter.

CASE 16 (III, 11; fig. 21).

This 12-year-old female cousin of the propositus was examined in New York on July 6, 1956, through the courtesy of Dr. Ramón Castroviejo, who also made the clinical records available.

Dr. Castroviejo first saw the patient in October, 1947, when she was three years old. She had a history of ectropion of all four eyelids since birth and slept with the eyes partly opened. Examination at this age showed slight ectropion of both upper lids and more pronounced ectropion of both lower lids. Although the patient did not cooperate in the examination, it was evident that she had some inturned lashes and that corneal nebulas were present in each eye.

Surgical correction of the ectropion by skin grafting was carried out on both right eyelids on September 2, 1948, and on both left eyelids on November 4, 1948. The immediate results were good, and when the child was examined in July, 1956, the ectropion was completely corrected, the cosmetic results were excellent, and the lids closed satisfactorily.

Examination at this time showed six aberrant lashes on the upper right lid, arising from the posterior border of the margin at the junction of the nasal and middle third. Four aberrant lashes were similarly located in the lower right lid. These lashes rubbed the eyeball when the patient looked down and to the left. On the left side, one inturned lash arose from the posterior border of the margin of the upper lid near the outer canthus, and another arose in the lower lid in the same position. Both of these lashes, especially the lower, rubbed against the eyeball. The aberrant lashes were all dark but seemed weaker and thinner than the lashes in the normal anterior rows.

The palpebral fissures measured 25 mm. horizontally and 12 mm. vertically in the right eye, with maximum effort. In the left eye the corresponding measurements were 24 mm. and 12 mm.

The patient had a dense, vascularized nebula in the lower third of each cornea. She had considerable photophobia.

REVIEW OF THE LITERATURE

ECTROPION

The 12 case reports which make up the literature on congenital ectropion can be divided into two groups. The first group (Group 1 of the proposed etiologic classification) consists of only five cases. The other seven cases, including the first case reported, by Adams[6] in 1896, are instances of group 2 of the proposed classification.

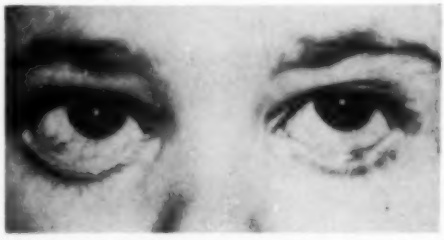

Fig. 21 (Picó). Ectropion of all eyelids, before surgical correction, slight in the upper lids and more marked in the lower lids (III, 11, Case 16).

Group 1. The first case of congenital ectropion of the eyelids was reported by Urmetzer[7] in 1914, at a meeting of the Vienna Ophthalmological Society, and was mentioned by Landau[8] in his 1947 contribution. The patient, whose age and sex are not mentioned in the abstract available to me, also presented abnormal shortness of the affected lower lids, severe redness of the lid margins, and madarosis. Complete closure of the lids was not possible. When the patient was asleep, the palpebral fissure remained open for a space of three mm. The lids were scarcely longer on stretch than when they were relaxed. Treatment consisted of the application of ointment and a bandage during the night; the results were not stated.

In 1915, Treacher Collins[9] reported congenital ectropion of the lower eyelids, present since birth, in a four-month-old child. Examination also revealed well-marked epicanthus and partial ptosis of the lids, which appeared too large to fit the eyeballs closely. There was no family history of congenital defects of the eyes or eyelids.

Operation was done when the child was four years and 10 months old; during the interval there had been no change in the anomaly. A wedge-shaped piece of tissue was resected through the whole thickness of the right lower lid, just internal to the outer canthus. Microscopic examination of the specimen showed the orbicularis to be well developed and the group of fibers near the free border of the lid (the muscle of Riolan) to be exceptionally numerous. The epithelium of the palpebral conjunctiva was several layers thick, and there was some papillae formation, apparently the result of exposure. There was no description of the tarsus.

In 1944 Gordon and Cragg[10] reported an instance of congenital ectropion, associated with bilateral ptosis, in a 23-year-old white man, who complained of ocular discomfort because of the deformity. He also suffered from mild epiphora. He slept with his eyes open, and a mucoid discharge accumulated in the palpebral fissures during the night. Dust and dirt produced so much irritation that he could work only in selected environments.

The palpebral fissures sloped slightly downward and outward. Mild ectropion was present in the outer half of all four lids, especially the lower lids. The upper lids were abnormally short and could not be elevated actively. The lashes were normal in position and distribution.

Treatment consisted of skin grafting followed, four months later, by the creation of fascial slings to secure the upper lids to the frontalis muscles and thus overcome the ptosis. The tarsus was not described in the notes on examination or operation.

The patient had no personal recollection of his father but had been told that he had suffered from a similar deformity of the eyelids. Two of the patient's six brothers and one of his three sisters also had similar defects, though less marked than his own. The mother's eyes were normal.

The cases reported by Landau[8] and Erdmann[11] are described under the heading of distichiasis, since the anomaly of the lashes was more pronounced in them than was the ectropion.

Group 2. The first recorded case of congenital eversion of the eyelids, reported by Adams[6] in 1896, concerned a two-month-old child who was born with both upper lids completely everted and greatly swollen. The palpebral conjunctiva was markedly thickened at the time of the examination. When the lids were placed in the normal position, they remained in it only while pressure was maintained. The use of compresses restored the lids to a permanently normal position in about a month.

In 1909, Erb[12] reported congenital ectropion of both upper eyelids in a two-day-old child, born after a prolonged labor due to breech presentation. The everted upper eyelids, which were large and edematous, were covered with bluish-red mucosa, which in turn was partly covered with scabs. Active bleeding followed the removal of the scabs.

The lids were replaced without difficulty after the child was anesthetized. They were held in position by adhesive tape. When it was removed five days later, they were in their normal position and the eyes opened well, but the upper eyelids were still larger than normal and were atonic.

In 1920, Mertens[13] reported congenital ectropion of both upper eyelids in a two-day-old child, born after a long and difficult labor due to a contracted pelvis. The upper eyelids were completely everted and the conjunctiva, which was enormously swollen, was puffy and red. Treatment consisted of the application of boric acid and vaseline.

The child was well developed and was apparently normal immediately after birth, but death occurred at four months from hydrocephalus and convulsions. The physician in charge of the case reported that the ectropion had been relieved, but that the palpebral fissures were narrowed, especially at the inner canthus, and were oblique from the upper outer toward the lower inner angles.

In 1954, Young[14] reported congenital ectropion of the upper eyelids in a newborn infant weighing seven pounds 12 ounces and born after an easy six-hour labor. The pregnancy had been entirely normal except for a mild pre-eclamptic toxemia in the seventh month. Five previous pregnancies had resulted in normal children. The sixth (immediately preceding) pregnancy had terminated in a stillborn child with hydrocephalus and congenital meningocele.

During the labor in question, as soon as the head appeared at the outlet, it was observed that both upper eyelids were completely everted and that their entire inner surfaces, together with a large part of each upper transitional fold, were exposed. The transitional folds were so swollen that the lid openings were completely obliterated. The ectropion appeared as large edematous swellings covered by bluish-red mucosa; when the child cried, the swelling increased and the coloration became more distinctly bluish. The lower lids appeared normal, but the eyes, which were observed only with great difficulty after retraction of the lower lids, seemed smaller than normal. The infant had the characteristic stigmas of mongolism.

On the eighth day of life, under general anesthesia, the excess tissue beneath the right upper eyelid was excised and the raw conjunctival margins were united by a continuous suture. The child died suddenly the following day, after an attack of cyanosis. Death was apparently unrelated to the operation.

Complete histologic examination of both upper lids at autopsy showed no intrinsic defects. A marked overdevelopment of the epicanthal fold was demonstrated on the outer surface of each upper eyelid, in the region of the inner canthus. Section of the eyeballs revealed all the intraocular tissues to be normal in structure and position.

In 1954, Ostriker and Lasky[4] reported a case of congenital ectropion, which, they stated, had no previous parallel in the literature, though the description and photographs suggest that this case closely resembles the cases reported by Erb,[12] Mertens,[13] and Young,[14] which have just been summarized. In Ostriker and Lasky's case, a newborn child, born after a short, spontaneously terminated labor, showed "thickened, swollen, injected, totally everted upper eyelids," without other associated ocular or general anomalies. By the third week of life the eyelids had become normal and so remained.

In 1955, Hopen[15] reported a case of complete congenital eversion of the upper eyelids in a full-term Negro boy, born after a labor which was uneventful except for a slight arrest of the head on the perineum. The mother, a 14-year-old primigravida, was RH positive.

The upper eyelids were markedly chemotic, thickened, and edematous and were totally everted. The condition was not improved by the continuous use of saline compresses, chlortetracycline (Aureomycin) ointment, and adhesive straps. On the fifth day of life, under light ether anesthesia, the lids on

both sides were sutured together in the outer, middle, and inner aspects of the margins with three mattress sutures of black silk applied over small rubber dams. Two days later, when the sutures were removed, the lids were in normal position, and apposition was satisfactory, even when the child cried. Improvement was permanent.

In 1955 Mazhar[16] reported congenital eversion of the upper eyelids in a seven-day-old Arab child, born at full term. The eversion, which was associated with chemosis and edema, had been present since birth. There was a moderate amount of secretion. Treatment by warm saline compresses and oxytetracycline (Terramycin) ointment restored the lids to normal position and condition within two weeks, so promptly, in fact, that the author wondered whether he had really been dealing with a true developmental anomaly.

Distichiasis

Distichiasis, a term derived from the Greek words meaning double and row, indicates the presence of an aberrant posterior row of eyelashes which emerge from the sites of the normal openings of the meibomian glands. The term was suggested by Fuchs[5] in 1889. Erdmann,[11] in 1904, championed the term distichiasis for the true congenital condition. At that time the terms trichiasis and distichiasis were still being used rather loosely, to indicate any sort of abnormal position of the eyelashes. DeVoe and Horwich[17] employed the term tetrastichiasis, apparently because in their case there were four rows of lashes, two normal and two aberrant, on the affected lids.

Distichiasis, which is a congenital condition, must be differentiated from the acquired abnormal condition known as trichiasis, a word which indicates malposition and maldirection of the lashes. Fuchs considered the noncongenital forms of distichiasis, such as occur in trauma, as basically the same as trichiasis, from which they differ only in degree.

The first report in the literature on congenital distichiasis, by Becker[18] in 1867, consisted of two cases. The original article is not available to me, and I quote from Kuhnt's summary of the cases.

The first patient, an eight-year-old girl, had suffered from disorders of the eyes for six years. The cause of the severe spasm of the lid was found to be a second row of cilia on the inner side of the lid margin, immediately adjacent to the orifices of the meibomian glands. These cilia were light blond and so regular in arrangement, length, and direction that when the lid was lifted from the eyeball, they looked like the teeth of a fine comb. When the lid lay upon the eyeball, the lashes adapted their form to its convexity; they brushed against the conjunctiva and cornea upon up and down movement, but without injury to them. The intermarginal portion of the outer border of the lids was wider than normal but had the peculiar shine of normal lids. In this area were a number of individual, extremely fine, scarcely visible small hairs.

Becker's second patient, a 13-year-old girl, suffered from catarrhal conjunctivitis. The only abnormality evident on examination was a second row of cilia on both lower lids, close to the openings of the meibomian glands. These lashes turned inward and irritated the eyeball.

In 1880, Nicati[19] reported a patient with photophobia and conjunctivitis caused by distichiasis. When he excised a piece of the lid margin, microscopic examination showed the points of implantation of the abnormal cilia, which were readily recognizable as they emerged from the inner edge of the lid margin, opposite the outer row of cilia. There was no doubt that these aberrant cilia emerged from the openings of the meibomian glands. Between the normal and anomalous rows of cilia the lid margin was entirely normal.

In 1891, Herrnheiser[20] reported a case of distichiasis in a 12-year-old boy who had suffered from red eyes throughout childhood.

There was a double row of cilia on all four lids, the second rows taking the place of the normal orifices of the meibomian glands. The aberrant cilia were lighter in color than the normal lashes and resembled the lanugolike hair the patient had over his whole body. The aberrant lashes pointed upward or downward, depending upon their location, and brushed against the eyeball, apparently without harming it.

Examination of serial sections of a piece of tissue excised from the lid margin showed that each aberrant eyelash corresponded in location to a gland whose excretory duct opened into the hair follicle. The roots of the cilia lay almost entirely outside of the excised section. The glands opened, like sebaceous glands, into the upper half of the hair follicles but had the structure of meibomian glands. They terminated within the excised portion and apparently did not extend upward. The number of both cilia and glands corresponded to the normal number of meibomian glands. Microscopically, no true meibomian glands were found, and the glands which were present appeared very slightly developed and seemed to serve as appendages to the abnormal cilia.

In 1899, Wood[21] reported two cases of congenital distichiasis, in a father and a daughter. Each patient had an aberrant row of fine white hairs which emerged from the inner border of the lid margin and at times brushed the cornea. Hotz,[22] in a discussion of this paper, mentioned a similar case, in which fine cilia were apparently implanted in the secretory ducts of the meibomian glands.

In 1899, Westhoff[23] reported distichiasis in an eight-year-old child who suffered from photophobia, left convergent strabismus, and reddening of the conjunctiva. On the sharp inner border of each eyelid were about 15 aberrant lashes which were normal in appearance except for their unusual length. On the upper lids these lashes emerged only from the inner border. On the lower lids a few also emerged at some distance from the border. The orifices of the meibomian glands were visible in this case. This child's anomaly had existed since birth. A similar condition was present in her mother, her three-year-old brother, and her mother's brother. Two other siblings were normal.

In an extensive paper on distichiasis published in 1899, Kuhnt[24] described a 52-year-old woman who had suffered from inflamed eyes and photophobia since childhood and who had required epilation for many years.

There was considerable blepharospasm and lacrimation on examination. The conjunctiva was slightly injected. On both corneas were dense and diffuse cicatricial opacities. Vision was limited to finger counting at 1.0 to 1.5 meters.

On each lid, in addition to the normal anterior rows of cilia, a row of fine, light, soft lanugolike lashes emerged directly from the intermarginal portion. These hairs, which were easily visible to the naked eye, were turned toward the eyeball and brushed against the cornea with each movement. By means of a magnifying glass it was seen that the intervals between them were similar to the intervals between the orifices of normal meibomian glands. The orifices of the normal glands were, however, not demonstrable.

On microscopic examination of a full-thickness wedge of tissue from each lid, the prominent findings were: absence of the meibomian glands; their replacement by well-developed cilia with all their characteristics; the presence of a double row of Krause's glands in the tarsus; and the presence of unusually well-developed Moll's glands at the posterior row of pseudocilia. The tarsus seemed less firm than normal.

This patient suffered from elephantiasis of the calves, as did one of her daughters, but no further evidences of congenital anomalies in the family could be elicited.

In 1904, Erdmann[11] reported three cases of congenital distichiasis in three generations of a family. The grandmother, who was 52 years of age, stated that her parents and sisters had had "weak eyes," and that she herself had suffered from inflamed eyes and

had required epilation since early childhood. There was slight photophobia in both eyes. The lid margins were moderately thickened. The lids were in normal position, though the two lower lacrimal puncta were slightly everted. The tarsal conjunctiva was red and swollen, and the palpebral conjunctiva was greatly injected. Both corneas showed considerable cloudiness and vascularization in the lower portions, and there was a small corneal ulcer on the left.

Examination with a magnifying glass showed rows of aberrant cilia on all four lids. These lashes, some of which were light and some pigmented, were soft, three to five mm. long, chiefly of the lanugo type, and without the thickness of the anterior cilia. They lay close to the rather sharp inner borders of the lid margins and parallel to them. They emerged from tiny, regularly arranged openings, which corresponded in number and position to the openings of the meibomian glands. Hairs appeared in the lower lids in all of these openings, in almost complete, regular rows, but many openings on the upper lids were without hairs. The aberrant lashes pointed upward or downward, depending upon their location. Sometimes they pointed backward, the longer lashes touching the eyeballs with their entire surface and the shorter lashes touching them only with their points. The aberrant lashes brushed against both the cornea and the conjunctiva.

Histologic examination of sections excised from the lower lid showed moderate thickening of the tarsus, whose tissue was less firm and less sharply delimited from the surrounding area than in a normal lid. In the tarsus, close to its transition into the tissue of the lid margin, were embedded glandular alveoli, which were scantier than normal meibomian glands but were quite similar to them in structure and position. These alveoli were connected with the hair follicles. The posterior cilia, which were quite rudimentary, were of small diameter. The papillae were small. The Moll's glands which were present opened into the hair follicles.

The daughter of this patient, who was 21 years of age, had had frequent inflammations of the eyes in childhood and what she described as occasional "strong flows" of tears later in life. She had distichiasis of all four lids, but the aberrant eyelashes were less numerous than those of her mother or her daughter. A large number of openings on the inner margin of the lids, which were assumed to be orifices of meibomian glands, were without lashes. The structure and position of the aberrant lashes were similar to these features in the mother's case.

This second patient's child, a six-year-old girl, had always had red eyes, associated with slight lacrimation. Examination showed irritation and photophobia. The tarsal conjunctiva was somewhat reddened and swollen and the conjunctiva of the eyeball was slightly injected. The cornea looked normal on superficial inspection, but on closer examination was found to have a slightly roughened surface and to be less shiny than usual, because of a slight irregularity of the epithelium.

On the inner border of the margin of all four lids, very fine, light, lanugotype hairs, six to eight mm. long, emerged from openings corresponding to those of the meibomian glands. These lashes, 25 to 30 in number on each lid, formed complete, perfectly regular rows. Although they turned slightly inward and brushed the cornea and conjunctiva with their entire surfaces, there was no evidence of any significant irritation.

Histologic study of a section from the right upper lid showed a few glandular acini which corresponded in all respects with meibomian glands except that they opened into well-developed hair follicles of weak lashes instead of into a common excretory duct.

In 1906 Brailey[25] reported a case of a 14-year-old boy with a chronic conjunctivitis which had caused hyperemia and excoriation of the lid margins. The cornea showed delicate maculas. Numerous fine hairs were present in both upper and lower lids; they rose from the posterior margins, presented

as single, closely placed rows, and were delicate and almost colorless. They were about half the length of the normal anterior cilia, from which they were separated by the full width of the intermarginal space. These aberrant lashes lay upon the cornea. On examination with a lens, it was found that what had at first been assumed to be the orifices of the meibomian glands were actually follicles of the accessory cilia; the normal glands were not present. Microscopic study of a small specimen obtained from the center of the upper lid, including the conjunctiva and the entire thickness of the tarsus, showed no trace of the meibomian glands or of any epithelial cells. The tarsus was of normal thickness, density, and structure. Brailey considered that the section was too small to permit conclusions, adding that glands might have been present in other parts of the tarsus or in a rudimentary state near the lid margin, no portion of which was included in this specimen. This patient had two accessory bicuspid teeth, representing another possible instance of developmental error of the epithelial structures.

In 1912 Traquair[26] reported distichiasis in an 18-year-old boy who had suffered from irritation in the left eye for about a year. The lower lids were slightly shortened vertically and the skin over them seemed less loose than usual. There was a minimal degree of distichiasis in both lower lids. No meibomian glands could be demonstrated and it could not be determined whether the tarsal cartilage was present or absent. The condition was considered to constitute an almost complete absence of cilia and intermarginal zones.

Three other siblings were normal, but a similar anomaly was present in the patient's father, without obvious distichiasis. "Small and tender" eyes were characteristic of the paternal side of the family; the paternal grandmother and two other relatives on that side had "small" eyes. It seemed reasonable according to Traquair, to explain the anomaly by the presence of an abnormality of the cilia.

In 1913, Begle[27] reported distichiasis in a 30-year-old woman who had suffered from irritation of the right eye for a year; a year earlier she had a corneal ulcer. The inner border of the margin of each lid presented a row of cilia, instead of the orifices of the meibomian glands. The aberrant cilia were black and three to five mm. long; because the angle of the margin was slightly rounded, they rubbed against the cornea lengthwise instead of with their tips. There were 13 and 17 accessory cilia, respectively, on the lower lids, and seven and eight, respectively, on the upper lids. A few glandular openings were observed on each lid in line with the accessory cilia. A mild degree of conjunctivitis was present, and there were several opacities and small corneal ulcers on the left eye.

Microscopic examination of strips of tissue from the lower lids, which included the upper third to the upper half of the tarsi, revealed absence of the meibomian glands and their replacement by (1) hyperplastic sebaceous glands which discharged into the follicles of well-developed accessory cilia and (2) small, simple, independent sebaceous glands scattered irregularly just beneath the lid margin and discharging upon its surface.

In 1923, von Szily[28] reported distichiasis in a 15-year-old boy, who complained of severe photophobia. His parents and siblings presented no anomalies. Since birth, this boy has had a second row of cilia on the inner border of each lid margin; they apparently took the place of the openings of the meibomian glands. There were 36 hairs all turned slightly inward, on the right upper lid; the few glandular orifices present were free of hair and were covered with secretion. All of the additional lashes brushed against the cornea and, when the eyes were closed, their points lay in the conjunctival sac. There were 30 aberrant cilia on the right lower lid, spaced at fairly regular intervals; they were finer and lighter than the normally located cilia, and their points turned inward. The 24 accessory cilia on the left upper lid were strongly developed and were only slightly

lighter than the normally placed anterior row. They appeared less frequently in the nasal third than elsewhere. When the lids were open, these cilia were chiefly bent outward, but when the lids were closed, they were also within the palpebral fissure. The cilia on the temporal half of the lid, whose tips were bent, were entirely within the conjunctival sac. On the left lower lid, 26 aberrant lashes all emerged from the orifices of the meibomian glands. The conjunctiva of the lower lids was thickened and swollen. There were several opacities in the lower right cornea.

Examination of sections of the posterior portions of both upper lids and the left lower lid, removed through an intermarginal incision, showed the tarsus of normal firmness and almost normal length and width. The ciliary follicles and hair shafts were normal. The structure of the sebaceous glands was similar to that of meibomian glands, but these glands were too small to be meibomian glands and too strong to be ordinary hair follicles. Moll's glands were absent in some areas, particularly in the lower lid; elsewhere they were of various degrees of development.

In 1924, Blatt[29] reported five instances of congenital distichiasis in three generations of a single family, all of whom suffered from nervous diseases. The grandmother, who had had "seizures" in childhood, had two rows of cilia on each of the lower eyelids. Her son, who had epilepsy, had two rows on all four lids. Her daughter, who also had epilepsy, presented the same anomaly. The elder son of the son, like his paternal grandmother, had two rows of cilia on the lower lids. His younger brother, then aged 17 years, was just beginning to complain of his eyes; he also had two rows of cilia on both lower lids. A daughter of the daughter, aged 12 years, had no ocular complaints and presented no ocular abnormalities. Microscopic examination of the cilia in some of these cases, showed structural abnormalities.

In 1927, Doherty[30] reported distichiasis in a seven-year-old boy, who had a second row of lashes arising from the inner margin of each lid. The accessory lashes were as numerous and as well developed as those of the normal anterior row. Although they rested against the cornea and the patient complained of lacrimation and of a feeling of sand in his eyes, no corneal opacities could be detected with the microscope. The aberrant lashes were removed by electrolytic epilation.

In 1927, Frolowa[31] presented the case of a 14-year-old boy who had suffered from lacrimation and photophobia since childhood. In place of the orifices of the meibomian glands he had 12 accessory lashes on the upper right lid, 15 on the lower right lid, and 15 and 14, respectively, on the left lids. Under the microscope, the aberrant cilia were lanugolike, being shorter, thinner, and lighter than normal cilia. The medulla was either absent or scarcely noticeable. The shafts of the lower cilia brushed against the cornea, and when the eyes were closed the cilia rested on the conjunctival sac. In some areas in the accessory posterior row in which lashes were absent were small openings, at distances which corresponded to the openings of the meibomian glands. The corneas were transparent.

Microscopic examination of serial sections obtained perpendicularly to the length of the tarsus showed that in some areas in the lower lid the acini of the meibomian glands were almost normal in number and size. In other areas the glands were rudimentary, particularly when there were hairs in the lumen; these glands were small, with two or three acini each, and suggested ordinary sebaceous glands. In some sections roots of the anterior row of normal cilia were seen next to the atypical lashes emerging from the meibomian glands. The roots of the anterior lashes were much thicker. Some sections from above the tarsal tissue showed a round, cystlike formation, with clusters of fine rolled hair.

This patient's father presented the same anomaly, except that it was confined to the lower lids. Other members of the family, including the deceased mother, were reported to be without anomalies.

In 1935, Halbertsma[33] reported distichiasis in a 37-year-old man who had complained of photophobia, tearing, and blepharospasm, more troublesome on the left, since the age of 18 years. No family history of eye difficulties was secured. There were double rows of cilia on all four lids. The cilia of the posterior rows were less thick and were located vertically, at irregular distances. The posterior hairs numbered from 10 to 20 on each row, and each row formed an angle of 90 degrees with the normal anterior row. The aberrant lashes were from two to three mm. long. Their shafts touched the bulbar conjunctiva as well as the cornea, which presented numerous opacities, especially on the left. The bulbar and tarsal conjunctivas were red and swollen.

Histologic examination of an excised specimen showed part of the shaft of a pseudoeyelash present in the orifice, which opened near the posterior border of the lid. The other end of the hair follicle formed the orifice of a glandular mass which appeared to originate from sebaceous glands and which consisted of some 10 to 12 small lobes. These small lobes united to form larger lobes, one of which opened directly in the hair follicle. On both sides of the follicle were small glandular lobes which, in the section, were of the same structure as the glandular mass; they were regarded as possible outgrowths. A single small gland, a third of the way from the free border of the lid and quite close to the stem, had no opening. Apart from the acinous gland described there was no glandular tissue at the point of emergence of the shaft.

In 1938, da Pozzo[23] reported two instances of congenital distichiasis, in a mother and one of her sons. The woman had begun to complain of lacrimation, photophobia, and the sensation of a foreign object in the eyes at the age of 27 years, but her husband had been aware of the abnormal growth of her eyelashes for the preceding seven years. There were seven accessory lashes on the right upper lid, two on the left upper lid, and eight on the left lower lid. The posterior lashes, whose shafts brushed against the cornea, were shorter and less pigmented than the anterior lashes. All the hairs emerged from follicular orifices in the normal location of the orifices of the meibomian glands. Slit-lamp examination revealed orifices of meibomian glands from which no hairs emerged. The conjunctiva was reddened.

Histologic examination of two pieces of tissue from the left lower eyelid revealed both normal and rudimentary meibomian glands. The latter were similar in structure to normal glands but presented only two or three acini.

The five-year-old son of this woman presented the same condition as the mother. The conjunctiva was not reddened and he did not complain of any discombort.

In 1947, Landau[8] described distichiasis in a 16-year-old boy who, when his eyes were closed, presented a bilateral lagophthalmos of about 2.5 mm. The height of the upper lids by Fuchs' technique was 24 mm., the vertical extension 30 mm., and the coefficient 1.24. In both upper lids were rows of well-developed accessory lashes. The cutaneous portion of the lid margin was slightly everted, particularly in the temporal portion. A fine dark line in the subtarsal sulcus, from which thin vertical streaks radiated, was thought to indicate the location of hair follicles of supernumerary cilia showing through the palpebral conjunctiva.

In 1949, Halberg and Paunessa[34] reported distichiasis associated with incomplete mandibulofacial dysostosis (Franceschetti's syndrome). There were double rows of lashes on all four lids, the accessory row on the upper lids being unusually long. Braley[35] reported a similar case in 1955, with severe corneal scarring.

In 1954 De Voe and Horwich[17] reported congenital entropion and distichiasis (which they called tetrastichiasis) associated with palpebral hyperpigmentation and mental deficiency in a 32-year-old Negro woman. An older sister stated that all these conditions

had been present since birth. The sister also presented tetrastichiasis and trichiasis and had a vestigial accessory finger on the lateral margin of each hand. Another sister also had polydactyly.

The patient presented entropion of the lateral half of each upper eyelid for a distance of 13 to 14 mm. The palpebral fissures were 27 mm. long. Tetrastichiasis was present for the full length of both upper eyelids and trichiasis for the lateral half. There was one row of meibomian orifices in each eyelid. Irritation of the cornea by the accessory cilia had produced scarring of the lateral third on the right, and of the lateral half on the left. On eversion of the eyelids, which was achieved with considerable difficulty, the normal concavity of the tarsal plates was seen to be exaggerated laterally in the vertical meridian.

When a Streatfield-Snellen operation was done on the left upper lid, the tarsus was found completely normal except for the accentuated concavity. The cut surface of the slim wedge which was removed was also entirely normal.

Comment

The literature of congenital ectropion

Congenital ectropion caused by hyperkeratosis or ichthyosis congenita, as well as the variety caused by congenital microphthalmia or orbital cysts, presents no etiologic difficulties. The other varieties (Groups 1 and 2 of my proposed classification) are somewhat more difficult to analyze from this standpoint.

There are five recorded cases in the first group, congenital ectropion due to absence or anomalous development of the tarsus. In Urmetzer's[7] case the description available is too slight to warrant any discussion of the etiology. In Erdmann's[11] and Landau's[8] cases, the chief emphasis is on distichiasis, there being only casual mention of eversion of the eyelids. The descriptions of both Collins's[9] and Gordon and Cragg's[10] cases suggest that they are comparable to some of the cases in my own series, though in neither, unfortunately, is the report sufficiently detailed for a complete comparison.

Although Collins's report includes laboratory examination of an excised specimen from the affected eyelids, there is no mention in it of the state of the tarsus. His original idea, that the anomaly could be attributed to a developmental defect of the muscle of Riolan, whose function is to keep the border of the lids in contact with the eyeball, was not borne out by findings; the muscle fibers, in fact, were unusually numerous. Collins's patient suffered from both ptosis and epicanthus, which were not evident in any of my series. Gordon and Cragg also do not describe the condition of the tarsus, although they could have examined it when skin grafts were applied to the lids. In their case the upper eyelids were short, as they were in Urmetzer's case and in several of my own cases. No abnormality of the lashes was found in either of these cases, in contrast to the distichiasis present with the ectropion in eight of my own cases.

The hereditary factor is not mentioned in Urmetzer's case. Collins specifically excluded it, though the possiblity exists, of course, that the reported case was the first in the family and that the defect might become manifest in future generations. In Gordon and Cragg's case the hereditary factor was evident. The father apparently had the same condition, as did two of six brothers and one of three sisters, though it was less marked in the siblings than in the original patient. The type of heredity suggests dominance, but one cannot be definite, since the history covers only two generations.

In the 10 living patients with ectropion in my series, the anomaly involved all four lids in four cases (Cases 9, 10, 13, and 16) and was complete in the lower lids but less marked in the upper lids in all the cases. One other patient (Case 1) had complete ectropion of both lower lids, with normal upper lids. The five other patients (Cases 5, 6, 7, 14, and 15) had only slight ectropion of both lower eyelids. One patient (Case 15) also had

a slight entropion of the left upper eyelid on upper gaze; the right upper lid was in normal position.

As already mentioned, no tarsus was found in two cases (Cases 1 and 5) on histologic examination of wedges of tissue removed from the lower lids; in Case 1 the ectropion was complete but in Case 5 it was only slight. These observations do not warrant generalizations concerning a definite relationship between the degree of ectropion and maldevelopment or some degree of weakness of the tarsus. On the other hand, the tarsal plates act as a skeleton for the lids, accounting for their shape and firmness,[36] and it is therefore reasonable to assume the ectropion is related to absence of the tarsus or to some degree of weakness in it.

The second group of cases of congenital ectropion takes the form of eversion of the upper eyelids. The eversion, which is observed immediately after birth and which disappears within days or weeks, apparently occurs during the passage of the child through the birth canal. It is not yet clear whether the associated edema of the palpebral conjunctiva is the result of the eversion, which itself is the result of birth trauma, or the eversion is secondary to a pre-existing edema.

Absence or deficiency of the tarsal plates was apparently not a factor in any case in this group, though it was specifically excluded only by Ostriker and Lasky[4] and by Young,[14] who reported the single histologic examination of the affected tissues. Only Mertens[13] mentioned an abnormal hereditary anlage as a possible factor, and he did not pursue the suggestion. There is nothing in the description of his case to support the idea except that the child presented characteristics of mongolism.

The literature of distichiasis

The literature of congenital distichiasis, which is far more extensive than the literature of congenital ectropion, can be discussed from several different points of view:

1. *Status of the tarsus, meibomian glands, and aberrant eyelashes.* Although Becker,[18] in the first recorded cases of distichiasis, noted that the aberrant cilia emerged immediately beside the orifices of the meibomian glands, Nicati,[19] in the second report of the anomaly (the third recorded case), showed by microscopic examination of an excised specimen that these lashes emerged from the orifices of these glands; he apparently studied only the points from which the aberrant lashes emerged. In his opinion, the meibomian glands were similar to sebaceous follicles, whose structure and function they shared.

Continued investigation of this phase of the anomaly produced widely different findings. In some cases the meibomian glands were rudimentary and undeveloped. Thus in Herrnheiser's[20] case, the first to be studied histologically, a posterior row of cilia in each eyelid replaced the orifices of these glands. The number of aberrant lashes corresponded to the number of glands. No true meibomian glands could be identified and the structures which apparently represented them were only slightly developed. Erdmann[11] found these glands poorly developed in two of his cases; lashes were present in some of the orifices but not in others.

In von Szily's[28] case, the posterior cilia emerged from the orifices of the meibomian glands. In his detailed histologic studies[37] he observed that these hairy glands were rudimentary. The tarsus was of normal firmness, length, and width. These findings are in contrast to Kuhnt's[24] impression that the component tarsal elements in his case were less firm and less closely textured than in cases in which normal meibomian glands are present.

In Landau's[8] case, as in Kuhnt's, hair follicles had apparently taken the place of these glands and occupied their ducts. In Brailey's[25] case, the orifices of the glands were replaced by the openings of follicles of the accessory cilia. Microscopic examination showed the tarsus to be of normal thickness, density, and structure, but there was no trace

of meibomian glands or epithelial cells, though the author, as already mentioned, felt that the small size of the specimen did not warrant conclusions.

In some cases, such as Traquair's,[26] there was no histologic examination but an apparent but not conclusive clinical absence of the meibomian glands. In other cases, such as that reported by De Voe and Horwich,[17] there was a row of orifices on each eyelid. In their case the tarsal plate felt normal except for accentuation of the concavity.

In some cases, such as Begle's,[27] a row of cilia on the inner border of the margin of each lid replaced the orifices of the meibomian glands. Histologic examination revealed the replacement of these glands by hyperplastic sebaceous glands, which discharged into the follicles of well-developed accessory cilia. Frolowa[31] found abnormal cilia replacing the orifices of the meibomian glands in the lid margins; when no hairs were present on the posterior border, either no openings were apparent or there were small openings at distances corresponding to the location of the meibomian glands.

In many of my personal cases the tarsus was either absent on palpation or was small and weak. Its complete absence in the lower lids was established in Cases 1 and 5, when operation was done for correction of an ectropion, and was further confirmed by histologic examination of excised specimens. Both of these patients had some aberrant cilia in the upper lids but none in the lower. It would have been interesting to determine histologically the relation of the posterior cilia in the upper lids to any meibomian glands present, but there was no justification for excision of tissue for examination. In both of these cases tarsal structures were clinically present in the upper lids, in which there was some distichiasis. This is not, however, conclusive evidence; a structure which is normal on clinical investigation may prove undeveloped or otherwise abnormal under the microscope.

The cilia differed widely in the number, appearance, and direction of their shafts. This was evident in my own cases as well as in the literature. Some patients had only a few additional lashes on one or two lids, in areas in which the orifices of the meibomian glands usually open. Others had almost complete accessory rows in all four lids. In Case 3, in which the distichiasis affected only the lower lids, orifices of the meibomian glands were absent, though they were present and apparently normal on the upper lids. No orifices were observed in the other nine cases of distichiasis, even when they were not replaced by aberrant cilia.

In most instances the hairs were pale, thin, and of the lanugo types, though in some cases they were dark and quite similar to the anterior normal row; this was particularly true in the accessory row of lashes in the left upper lid in Case 15.

In most cases some of the posterior lashes rubbed against the cornea and bulbar conjunctiva, producing conjunctivitis, keratitis, and corneal scars. In other cases the hairs were inclined downward along the margin of the affected lid. In Case 15, the aberrant lashes assumed the general direction of the normal anterior lashes and did not touch the eyeball.

2. *Combined ectropion and distichiasis.* The combination of congenital ectropion and distichiasis was first reported by Erdmann.[11] In this case the lids were in normal position but the lower lacrimal points were slightly everted. In Landau's[8] case there was vertical shortening of the upper eyelids, the cutaneous portions of the margins being slightly everted.

The series of cases in the same family which I am reporting show an apparently unique combination of ectropion and distichiasis. Ten of the affected patients whom I examined personally had distichiasis, which in eight cases was accompanied by ectropion. The ectropion varied from a slight condition affecting only the lower lids to a very marked condition which in two cases affected all four lids. Information obtained about two other

members of the family from the surgeon who had operated on them indicated that ectropion was present in all four lids; no information was secured about distichiasis. Another deceased member of the family was reported to have had complete ectropion in all four lids, but distichiasis was not mentioned.

3. *Related anomalies.* In reported cases of distichiasis the related anomalies took several forms:

"Small and tender" eyes were mentioned by Traquair[26] as a feature of the ocular anomalies on the paternal side of his patient's family. In my own series, all 10 members of the family whom I have examined personally had short palpebral fissures horizontally, this anatomic characteristic giving the impression that the eyes were small. The grandmother of the propositus (I, 2) was reported by her sons to have had such eyes, though she apparently had neither ectropion nor distichiasis.

Vertical shortening of the eyelids was present in some of my cases, as in Landau's,[8] in which lagophthalmos and slight ectropion of the upper lids accompanied distichiasis. In his report, made in 1947, he revived Fuchs'[18] study, published in 1889, on the relation between the height and vertical extension of the upper eyelids, which Fuchs expressed as a coefficient. With a coefficient below 1.5 (1.9 in older persons), normal closure of the lids is impossible.

In all six of my own cases of distichiasis in which these measurements were made, the coefficient was lower than normal. In three other cases (Case 10, 13, and 16), no measurements are available, but the fact that the surgeons who operated on these patients found it necessary in each case to use skin grafts to correct the ectropion of all four lids suggests that the lids were abnormally short. In the two cases in which I operated personally (Cases 1 and 5), although the coefficient indicated that the lids were abnormally short, the ectropion present in the lower lids did not seem to be due to any lack of skin but rather to a relaxation produced by the absence of the tarsus. For this reason, the Kuhnt-Szymanowski technique was used in each instance.

4. *Other ectodermal defects.* Braley[35] and Halberg and Paunessa[34] reported mandibulofacial dysostosis, a syndrome caused by congenital arrest of the primitive ectoderm,[35] in association with distichiasis. The two accessory biscuspid teeth present in Brailey's[25] case are also evidence of an ectodermal defect. In Case 2 of my own series, in which distichiasis was present without ectropion, the patient had been operated on in childhood for congenital cataracts. In Case 4 the eyes were normal, but the child was a moron, possibly because of some ectodermal cerebral defect.

5. *Anatomic defects producing ectropion and distichiasis.* Ectropion and distichiasis apparently have some anatomic defect as their common background. This is suggested by some of my own cases, in which the same patient presented both anomalies, sometimes in different lids and sometimes in the same lid. The cause of both conditions seems to be a defect of tarsal development. Vertical shortness of the eyelids is probably also due to a deficient development of all the tissues in the lids secondary to primary absence or deficiency of the tarsus.

Two chief theories have been advanced to explain the occurrence of distichiasis. Kuhnt,[24] arguing from histologic evidence, concluded that it is a heterotopic abnormality in which the meibomian glands are replaced by true cilia with characteristic adnexa. Brailey[25] concurred. Erdmann,[11] who, like Herrnheiser,[20] found poorly developed meibomian glands in his specimen, took issue with Kuhnt. His assumption was that in distichiasis, the anlage in which both meibomian glands and cilia develop does not differentiate, with the result that the development of the acini of the meibomian glands is rudimentary and a rudimentary eyelash also develops.

The second theory, that distichiasis may be

an atavistic phenomenon in which glands of a highly modified type fail to develop and are replaced by similar structures which are more primitive phylogenetically, was first advanced by Begle.[27] In his case, meibomian glands were replaced by hyperplastic sebacceous glands discharging into the follicles of well-developed accessory cilia. In von Szily's[37] opinion, in which Frolowa[31] concurred, congenital distichiasis is merely a return, in part, to a phylogenetically earlier form. Both anatomic and morphologic studies indicated to him that this anomaly is a true idotypical recessive hereditary deformity; that the posterior row of cilia is not merely a displaced row but one which has the characteristics of rudimentary hairy meibomian glands; and that these glands most probably represent phylogenetically intermediate stages of the series of transformations through which ordinary hairs have passed in the ancestors of present day mammals in their development into meibomian glands.

Blatt[29] also accepted the atavistic theory of origin. It is possible, he argued, that some disorder in the germinal layer may inhibit development of the gland, with the result, since the primary germinal layer is the same for both glands and hair, that hair is formed. This argument is in line with the general opinion that disordered development is determined in the germinal layer in congenital distichiasis because of its familial occurrence and because it is assumed that true normal meibomian glands originate from the hair anlage of the tegument of the lid margin. Like von Szily, Blatt regarded hairy glands as a physiologically lower type which was replaced, in course of time, by the higher type normal meibomian glands.

Frolowa's[31] findings may explain the different histologic data reported by various observers; in her case she found not only rudimentary glands with cilia, some of which resembled ordinary sebaceous glands, but also glands with the structure of normal meibomian glands. It is quite possible that the different histologic findings which have been reported by different observers may be explained, on this basis, as regressions to any one of several different stages, from true cilia, through rudimentary hairy glands, to true meibomian glands. da Posso[33] also found both normal and rudimentary meibomian glands in one of his cases.

The chief objection to the atavistic theory of human aberrations is that modern embryologists are extremely loath to accept it. On the contrary, they lean toward the belief that such changes are either true or specific mutations.

6. *Hereditary influence and mode of inheritance.* The possible hereditary factor in distichiasis was first mentioned by Wood,[21] whose two cases occurred in a father and daughter. Westhoff[23] also reported the condition in two generations (the patient, his mother, his brother, and his maternal uncle). In Frolowa's[31] case the condition was present in a father and his only son. da Pozzo[33] reported the condition in a mother and one of her sons, and in the case reported by De Voe and Horwich,[17] the condition was present in the patient's sister.

Erdmann[11] first reported distichiasis in three generations (grandmother, mother, and daughter; the anomaly was not present in two siblings in the third generation). Blatt's[29] contribution is particularly important from the standpoint of the hereditary factor: Five members of three generations of the same family were affected (the grandmother, a son, a daughter, and two sons of the son); a daughter of the daughter was not affected. These five cases indicate autosomal dominant inheritance. They are the first cases to be reported showing the affection in both sexes in three generations, although those affected in the third generation consisted of two sons of an affected son.

Waardenburg[38] some years ago expressed the opinion that hereditary findings in the cases of distichiasis reported in the literature are suggestive of dominance because of the true rarity of the condition, which he thought might possibly be polyhydridism.

The pedigree (fig. 1) of the family which I have studied gives evidence of a dominant autosomal inheritance of both ectropion and distichiasis, there being direct transmission of both conditions over three generations. The possibility of sex linkage of the pathogenic gene is eliminated by the absence of predilection for either sex in any of the generations. If there were sex-linked dominant X chromosomal inheritance, none of the sons of the affected fathers would show the condition, as some of them do.

The affected parents' genotype for the pathogenic gene is apparently heterozygous. In the first branch of the family, while the father was affected, 50 percent of his offspring were not. The large number of offspring in this family provided the opportunity to observe that four of the affected parents in the three generations had affected sons and daughters but that three of the four also had normal sons or daughters.

The pathogenic gene in this series of cases showed a high degree of penetrance. This is evident in the fact that more than half of the members of the family presented one defect or the other, or both. The expression of the pathogenic gene is somewhat variable, because the degree of ectropion or distichiasis shows considerable variance. There is no doubt of the expression, however, since of the 10 patients examined, only two showed distichiasis alone. The other eight showed both ectropion and distichiasis.

Summary

This thesis is based on the investigation of a family of 18 members in three generations, 11 of whom suffered from congenital ectropion, in eight instances associated with distichiasis. Two had distichiasis alone, and only five had normal eyes.

Ectropion and distichiasis are very rare congenital anomalies of the lids, as is shown by an extensive review of the literature. Previously reported cases of ectropion, numbering five at most, lack many of the characteristics of the cases in this family. The association of ectropion and distichiasis, reported in two cases at most, also lacks many of the characteristics of the cases in this family.

Histologic examination of specimens of two cases in this family showed absence of meibomian glands and either absence of the tarsal plates or the presence of only vestigal structures. This etiologic factor was not determined in the cases of ectropion previously reported. Absence, weakness, or some other deficiency of the tarsus was determined by palpation in the other affected cases, but such an examination cannot be regarded as conclusive.

Other characteristic features of this syndrome of congenital ectropion and distichiasis included narrowing of the palpebral fissures horizontally (described by the patients as "small eyes") and vertical shortness of the lids.

The mode of inheritance of this congenital syndrome in the family under investigation was evidently of an autosomal dominant type.

Ave Ponce de Leon 654.

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Ophthalmic Miniature

A. v. Graefe tried to refer all the symptoms of glaucoma to pressure, and under these circumstances, if an excavation was present, it was but natural to assume that its depth depended on the degree of pressure present. We now know that an excavation is an indication of disturbance of nutrition (of the optic nerve) and that the degree of pressure has but very little to do with its formation.

A. Mooren, *Archives of Ophthalmology*, **13**:15, 1884.

NOTES, CASES, INSTRUMENTS

NEW SCISSORS FOR ENLARGING CATARACT INCISIONS*

RAMÓN CASTROVIEJO, M.D.
New York

At the 57th annual session of the American Academy of Ophthalmology and Otolaryngology held October, 1952, I reported a new pair of scissors for enlarging cataract incisions. These scissors had spring handles, and the blades were of equal length and curved to conform to the curve of the limbus so that the incision could be accurately enlarged after the anterior chamber had been entered either with a keratome or a knife. These scissors were an improvement over previous models designed for similar purposes, but the blade which was introduced into the anterior chamber sometimes traumatized the iris or endothelium of the cornea.

For some years now I have been using a modification of these scissors† which has proved useful (fig. 1). The modification is as follows:

The inner blade, which is introduced into the anterior chamber, is one-mm. longer than the outer, or upper, blade, is rounded in the manner of a spatula, and the point is blunt in order to minimize trauma to the iris and endothelium. The second advantage of the longer inner blade is that it does not need to be removed from the eye upon each closure of the blade, so that, unless he so desires, the surgeon does not need to re-introduce the blade after each cut.

9 East 91st Street (28).

Fig. 1 (Castroviejo). New scissors for enlarging cataract incision showing that the blade which is introduced into the anterior chamber is spatulated and is longer than the outer blade.

* From the Department of Ophthalmology of St. Vincent's Hospital and the New York Eye and Ear Infirmary and New York University Post-Graduate Medical School.

† This instrument is manufactured by Storz Instrument Company, 4570 Audubon Avenue, St. Louis 10, Missouri.

A PRACTICAL EYE-DRESSING TRAY*

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Birmingham, Alabama

I am sure others have been through my exasperating experience with hospital eye dressing trays stacked with a small pan two or three inches in depth, bottles, ointment tubes, eye patches, gauze, and so forth, making efficient and reasonably sterile bedside treatment impossible.

An instrument company was kind enough to make up a tray which over a year's time has been purchased by hospitals in my area and has proven most satisfactory.

Figure 1 shows the individual receptacles for ointments, eye-dropper bottles, light, irrigation bottle, individual receptacles for cotton balls and eye-pads, and extra space for gauze, cotton applicators, and bandages. The tray is of one-piece stainless steel construction, with a convenient handle. The manner of construction has made it easy to

* The tray is available at the Storz Instrument Company, Saint Louis, Missouri.

Fig. 1 (McCoy). A practical eye-dressing tray.

identify and use medicines and equipment necessary for bedside care. Maintenance of sterile ointments and solutions is still a hospital problem. It depends upon the type of drop container employed, or the discarding of small amounts of solutions and sterilization of articles after use. Regardless of which system is used, this tray provides easy identification of used materials.

The adaptability, efficiency, and low price of this dressing tray are its chief assets.

501 Woodward Building.

SYPHILITIC TARSITIS*

A CASE REPORT

LARRY TURNER, M.D.
Durham, North Carolina

INTRODUCTION

One of the rarest manifestations of syphilis is involvement of the eyelids. Syphilitic tarsitis, generally regarded as a tertiary lesion, is believed to be rarest of all. Vancea[1] estimated that the cases of syphilitic tarsitis reported up to 1927 numbered only 25. According to Walsh,[2] there was only one case of syphilitic tarsitis in the records of The Johns Hopkins Hospital and, in that case, the diagnosis was not established beyond question. Cassady,[3] in reporting on gumma of the eyelid, summarized the literature on this subject up to 1950. He did not elaborate on syphilitic tarsitis. Although this condition was mentioned in older textbooks[4,5] of ophthalmology, most recent textbooks have given little space to this form of syphilis. Such authorities as Bull,[6] Fuchs,[4] and deSchweinitz[5] considered syphilitic tarsitis to be a gummatous lesion of the tarsus.

According to Duke-Elder,[7] tarsitis may accompany granular syphilitic conjunctivitis with involvement of the preauricular glands in secondary syphilis. In syphilis of the conjunctiva, an ulcer has been noted to invade the tarsus. Igersheimer[8] states that, in syphilitic disease of the lids and conjunctiva, one often has difficulty deciding whether a particular process belongs to the second or third stage of the infection. He is of the opinion that other findings of early syphilis, together with the presence or absence of necrosis and the ease with which spirochetes can be demonstrated, help one decide whether tarsitis is a manifestation of early or late syphilis. Tarsitis has not been generally regarded as a lesion of early syphilis, and, when it does occur, it is usually secondary to some other

* From the McPherson Hospital.

involvement of the lid or conjunctiva. The reason for reporting this case is that it shows primary syphilitic involvement of the tarsus. However, the history, laboratory, and pathologic findings represent an early stage of syphilis rather than the tertiary stage which is usually associated with this condition.

Case report

B. S., a 33-year-old married Negress, was referred to McPherson Hospital December 4, 1956, because of marked swelling of the right lower eyelid of approximately one month's duration. She stated that she first noticed a small "pealike" enlargement of the right lower lid which rapidly progressed in size. She consulted her family physician who gave her some tablets to take by mouth and prescribed a salve which she instilled into the eye. There was no improvement in the eye condition, and she was referred to the local ophthalmologist, who thought that the swelling of the lid had the appearance of a large chalazion. He incised the conjunctival surface of the lid and found it "hard and fibrous." He was unable to obtain any material by curetting. The patient was observed for two weeks and during this time the swelling increased to involve nearly the entire lid. It was at this time that the patient was referred to McPherson Hospital.

The family history and marital history were contributory in that the patient had had nine pregnancies with eight living children. There was one miscarriage. The last pregnancy, 10 months before, resulted in a normal, live, full-term male infant. The patient stated that blood tests during all of her pregnancies were negative, the last being reported in February, 1956.

Eye examination on admission to the hospital was not remarkable except for the markedly diffuse enlargement of the right lower lid. There was no discoloration or inflammation, either of the skin or the palpebral conjunctiva. Palpation revealed the lid to be hard but not uncomfortable. Difficulty was experienced everting the lid. It was noted that the right preauricular node was slightly enlarged but was nontender. The remainder of the physical examination was essentially negative.

Due to the history and nature of the growth, the right lower lid was explored through a skin incision. A pale yellow, tremendously thickened, avascular, rubbery tarsal structure was found. A biopsy was taken and the skin incision closed. Two days later, a VDRL test for syphilis drawn at the time of admission to the hospital was reported as positive in 32 dilutions. A repeat VDRL serology was again reported positive in 64 dilutions. The biopsy was reported by Dr. J. U. Gunter as follows:

Microscopic: This unusual lesion shows fibrosis, epitheliod cell formation, and dense infiltration with plasma cells, lymphocytes, eosinophils, and neutrophils. Plasma cells are quite conspicuous. The walls of some of the vessels contained within the lesion are fibrosed, thickened, and infiltrated with leukocytes. Occasional multinucleated cells are seen, but there are no typical giant cells.

Diagnosis: Chronic granulomatous inflammation of eyelid compatible with syphilis.

A Dieterle strain was then made of the tissue. This was reported as showing numerous spirochetes throughout the section.

When confronted with this evidence, the patient admitted that, eight weeks before her eye symptoms began, she had had extramarital relations with the same partner on three occasions. She denied having had any previous extramarital relations. She also recalled having had a slight rash about the right ear and neck for a few days prior to the onset of swelling of the right lower lid.

The patient was discharged to be treated for syphilis at a public health clinic near her home. When seen two weeks later, the enlargement of the right lower eyelid had completely subsided.

Discussion

Four different types of syphilitic tarsitis have been described. Le Roux[9] describes the first three types originally presented by Morax:

1. The marginal type is characterized by ulceration leading to deformity of the lid margin with complete or partial destruction of the lashes.

2. The nodular type is characterized by circumscribed thickening followed by deep gummatous ulceration. Before necrosis takes place, this may resemble a chalazion. The progress is always toward the skin, leaving the conjunctival surface normal. Whiting[10] reported a case which resembles this type.

3. The diffusely infiltrated type is characterized by diffuse painless swelling of the tarsus of one or more of the eyelids. Fuchs[4] states that this is seen as a primary involvement of the tarsus. He considered this to be due to gummatous infiltration. The swelling of the lid persists for several weeks and slowly disappears. He observed it was several months before the tarsus returned to normal size, or to less than its original size due to atrophy. Khalil[11] reported an interesting case of this type. Bull[6] states this type of tarsitis may be associated with other ocular findings of syphilis.

4. Vancea[1] has described a vegetative or

polypoid type. It began with swelling of the left upper eyelid. This was followed by a small, rose-colored, painless and slightly pedunculated tumor, like an irregular mushroom, which arose from the conjunctival side of the tarsus and resembled a meibomian epithelioma.

Although closely resembling type 3 in clinical appearance, the case reported here must represent an early stage of syphilis rather than a tertiary stage. The lesion of the right lower eyelid was the only manifestation of syphilis in this patient on admission to the hospital. A negative blood serology report 10 months before was confirmed by the hospital in which she delivered her last baby. According to the patient, the lid lesion appeared approximately eight weeks after exposure. Heckel and Beinhauer[12] state that gumma ordinarily appear from three to 20 years after chancre; however, gumma of the eyelids may develop from nine to 12 months after the primary lesion. In this case, there was also absence of marked necrosis. A special stain of the tarsus revealed numerous spirochetes which are difficult to demonstrate in gumma. A second VDRL showed an increasing titer. Considering these findings, it is possible that this lesion could be a chancre. According to Stokes,[13] there is no sharp demarcation between primary and secondary syphilis immunologically or clinically. He also states that chancre may assume any conceivable morphologic form.

When first observed, the lid lesion was described as having the appearance of a chalazion. When seen by this observer, the entire right lower eyelid was diffusely enlarged.

Fig. 1 (Turner). Note diffuse swelling of the right lower eyelid.

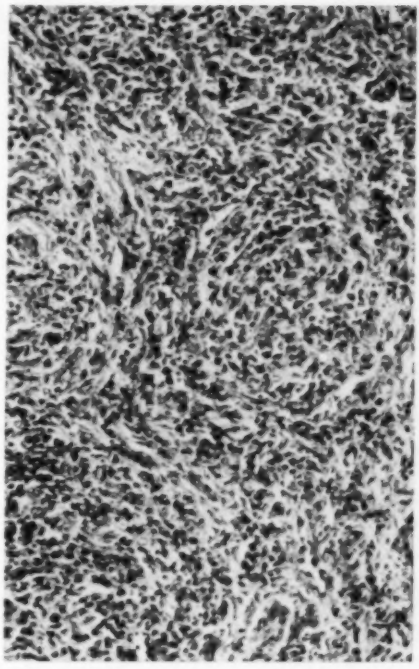

Fig. 2 (Turner). (Hematoxylin-eosin, ×162.) This photomicrograph shows a granulomatous inflammatory process with fibrous tissue proliferation, endothelial swelling, and diffuse infiltration with a variety of inflammatory cells, including some multinucleated cells.

Figure 1 shows the gross appearance of the right lower eyelid on admission to the hospital. Figure 2 is a low-power photomicrograph of a section stained with an hematoxylin and eosin stain. Figure 3 is an oil immersion photomicrograph of a Dieterle stain to show Spirocheta pallida.

Although syphilis is not as prevalent now as it has been in the past, tarsitis due to this disease has to be differentiated from chalazion, tuberculosis, and meibomian epithelioma. History and physical examination, together with serologic studies and pathologic examination, will help make this distinction. In tertiary lesions, serologic test may be positive in only approximately two thirds of the cases. According to Walsh,[2] and Heckel and Beinhauer,[12] it may be necessary to do a

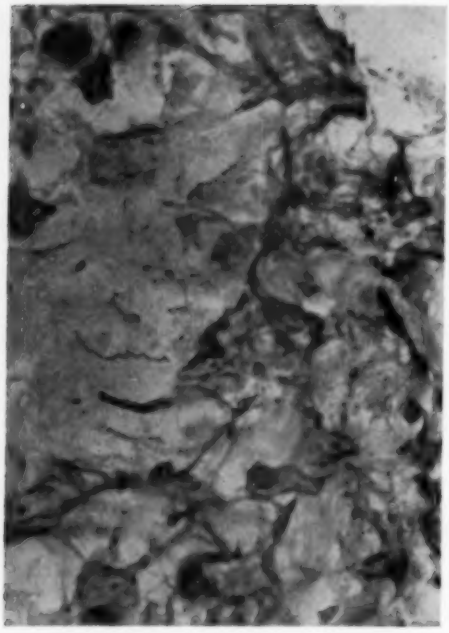

Fig. 3 (Turner). (Dieterle strain, ×1,788, oil immersion.) A typical spirochete is seen near the center of the picture and several others are slightly out of focus.

therapeutic test in order to make a definite diagnosis of late syphilis of the eyelid.

Conclusions

A case of syphilitic tarsitis is presented. Although generally regarded as a tertiary lesion, it is felt that this case represents an early lesion by history, serology, and pathologic findings. However, its gross appearance was quite similar to that described in tertiary syphilis.

Syphilitic tarsitis, though rare, is important as its varying manifestations may cause it to be confused with other lesions of the eyelid.

1110 West Main Street.

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HEMORRHAGIC GLAUCOMA*

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The term "hemorrhagic glaucoma" is applied to a group of glaucomas which have a common symptomatology and a varied etiology. The clinical picture is that of an intractable glaucoma. The eye is extremely painful, and shows high tension, severe congestion, marked corneal edema, and hyphema. This clinical picture is found as a complication of rubeosis of the iris, central retinal vein thrombosis, severe vascular retinopathies, and some malignant intraocular

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tumors. The exact mechanism responsible for the elevation of tension has not been well understood. Clinically, these eyes appear to have severe venous stasis, probably due to obstruction of the venous return circulation from the eye. The anterior segment of the eye is congested, the anterior ciliary veins are dilated and engorged, and the canal of Schlemm is filled with blood. One gets the impression that aqueous outflow is completely blocked, thus raising the intraocular pressure to very high levels.

This type of glaucoma is difficult to treat and there is little choice between retrobulbar injection of alcohol and enucleation in order to relieve the patient of his suffering. A filtering operation at the congestive stage causes severe postoperative bleeding and eventual loss of the eye. Cyclodiathermy has been disappointing.

These cases are emergencies in which the patient has to be relieved of excruciating pain as soon as possible. Recently I observed two cases of hemorrhagic glaucoma in which I succeeded in obtaining a white painless eye, although the tension remained very high and vision could not be saved.

Case reports

Case 1

W. H., a 72-year-old Negro with long-standing diabetes, was seen in February, 1956. He had a painful right eye of several weeks' duration. The eye was congested and extremely tender to the touch. The cornea was edematous and the tension was 85 mm. Hg (Schiøtz). After clearing the cornea with glycerine, it was noted that there was neovascularization of the iris in the area of the sphincter and at the periphery. There were a few posterior synechias in the lower part of the pupil, pronounced aqueous flare and a two-mm. hyphema. The anterior chamber was deep.

Gonioscopic examination showed that the angle was wide and open. The trabecular band was pigmented in the area of the line of Schwalbe. The canal of Schlemm was full of blood in the upper and lateral quadrants. In the lower quadrant, observation of the angle was impossible because of the hyphema. Vision in the right eye was reduced to light perception.

The left eye had 20/400 vision, not correctible because of severe retinitis proliferans. Tension was 22 mm. Hg (Schiøtz).

The patient was given a retrobulbar injection of one cc. of procaine (two percent) and one cc. of priscoline (25 mg.). The pain subsided within five minutes and the eye remained painless with the administration of 2.5-percent hydrocortone ophthalmic suspension instilled in the right eye every hour. Pilocarpine was not given. The patient also received 1,000 mg. of Diamox, followed by 250 mg. every six hours for several days. When the patient was seen two days later, the right eye was much less congested, corneal edema was less marked but the tension remained over 80 mm. Hg. The eye was less tender and the hyphema was a little smaller.

The patient was given a second retrobulbar injection of procaine and priscoline (one cc. of each). He was seen again one week later when the right eye was white and painless but the tension remained over 80 mm. Hg; the hyphema disappeared, the cornea was clear, and there were fewer blood vessels on the surface of the iris. Vision was reduced to light perception. The eye has remained hard and painless for the past two years.

Case 2

Mrs. T. L., a 67-year-old white woman, had a history of diabetes for the past 10 years. She was controlled with a diet and Orinase tablets. She had been checked periodically by an ophthalmologist who diagnosed mild diabetic retinopathy in both eyes. On November 22, 1957, she developed severe pain in the left eye and was seen by the resident who found a tension of over 80 mm. Hg (Schiøtz). He ordered pilocarpine (two-percent andeserine (0.5-percent solution) every half hour for two days. She also received 1,000 mg. Diamox followed by 250 mg. every eight hours.

When seen by me on November 30th, the patient had excruciating pain in the left eye. There was marked corneal edema and the tension was over 80 mm. Hg. After clearing the cornea with glycerine, a hyphema of two mm. was noted; there were also dilated capillaries in the lower area of the sphincter and two posterior synechias at the 6-o'clock position. The surface of the iris was muddy and there was an aqueous flare. On the nasal side there was a brushlike streak of clotted blood on the surface of the iris extending from the angle at the 10-o'clock position to the periphery of the iris at the 8-o'clock position. There was no rubeosis of the iris.

The gonioscopic examination showed a wide angle and a deep anterior chamber. The canal of Schlemm was full of blood. At the 10-o'clock position the blood broke through the internal wall of the canal and filtered across the intertrabecular spaces into the anterior chamber, giving rise to the hyphema.

The patient was given a retrobulbar injection of one cc. of procaine (two percent) and one cc. of priscoline. The pain in the left eye subsided within five minutes. The patient was told to discontinue the use of pilocarpine and eserine, was given a suspension of 2.5-percent of hydrocortone to be used every hour in the left eye. She continued to use Diamox (250 mg.) twice daily.

When seen three days later, there was little congestion in the left eye and the pain was very slight. Corneal edema had cleared considerably and there was little flare. The blood on the surface of the iris had absorbed and the hyphema was smaller. Tension was 80 mm. Hg and the canal of Schlemm remained engorged with blood. A second retrobulbar injection of procaine-priscoline was given and the hydrocortone and Diamox therapy was continued. On December 6th, the left eye was white and there was no pain, although the tension remained over 80 mm. Hg. The cornea was clear and funduscopy revealed mild diabetic retinopathy and a normal disc. Gonioscopy showed that the canal of Schlemm was still full of blood and the hyphema had almost disappeared.

On January 2, 1958, the condition of the left eye remained unchanged. The eye was white and painless, tension was over 80 mm. Hg, and vision was reduced to hand movements. The right eye had normal tension.

Comment

These two cases had many features in common. Both presented an acute attack of congestive glaucoma in an eye with a deep chamber and wide angle. The exact mechanism responsible for the acute elevation of the intraocular pressure is not well understood in these two cases. Certainly, it is not the same mechanism as in an acute congestive attack of narrow-angle glaucoma, where the pressure rises because of closure of the angle. In my two cases the angle remained open and there was no visible obstruction to outflow.

The course of events in these two cases can be reconstructed as follows:

The patients developed an acute iritis as evidenced by the presence of posterior synechias and aqueous flare. The congestion caused by the iritis appears to have resulted in obstruction of the return venous circulation from the eye, engorgement of the canal of Schlemm with blood, and complete arrest of aqueous outflow. Once the tension became elevated the venous return circulation became even more compressed and the congestion of the eye increased.

The presence of abnormal aqueous may well have been responsible for the pronounced corneal edema. Abnormal aqueous causes damage to the endothelium of Descemet's membrane and hydration of the cornea. It is possible that corneal edema is related more to the diseased state of the endothelium than to the high tension. There are many clinical facts on which this supposition may be based. In endothelial dystrophy of the cornea there is corneal edema in the presence of normal tension. In noncongestive angle-closure glaucoma the tension may go up to 70 mm. Hg but there is no corneal edema as long as the endothelium is intact. On the other hand, in iridocyclitis in which the endothelium is damaged by abnormal aqueous, corneal edema appears with tensions in the middle thirties. That the endothelium is damaged in these cases is evident from the keratic precipitates and pigment which become adherent to the diseased endothelial cells. In my two cases the corneal edema cleared when congestion and aqueous flare cleared. In spite of the persistence of tensions of over 80 mm. Hg for many weeks the cornea remained clear.

Pain and tenderness of the eye were probably due to the congestion and the inflammatory process caused by the iridocyclitis rather than to the high intraocular pressure alone. When the iritis cleared, pain disappeared although the tension remained very high.

This type of glaucoma should not be treated with miotics. In the presence of an active iritis, frequent instillations of miotics increase ciliary spasm and venous stasis. I found that short-acting mydriatics and intensive topical steroid therapy control the iritis promptly and thus relieve corneal edema and pain. In the presence of a wide angle there is no danger in dilating the pupil despite the high ocular tension.

It is difficult to explain why an iritis caused such a severe attack of high intraocular pressure. This is apt to occur in eyes predisposed to hemorrhagic glaucoma by some pre-existing condition such as rubeosis of the iris, retinitis proliferans, and central retinal vein thrombosis. Although the congestive attack is not always triggered by an

iritis, it is important to keep this possibility in mind. Failure to recognize the presence of an iritis may cause unnecessary suffering to a patient when he is given strong miotics in a vain effort to reduce the high intraocular pressure. This point assumes even greater importance when one remembers that it is difficult to diagnose an iritis in the presence of severe corneal edema. After clearing the cornea with glycerine, a flare and posterior synechias can readily be observed.

The role of engorgement of the canal of Schlemm with blood and its relation to the high intraocular pressure are worthy of some speculation. Blood in the canal of Schlemm is usually seen in hypotonic eyes where there is a reversal in the pressure gradient and a flow of blood from the anterior ciliary veins into the canal of Schlemm. Similarly, when pressure is exerted by means of the Koeppe contact lens on the perilimbal circulation, blood may be observed in the canal of Schlemm of normotensive eyes, but seldom in glaucomatous eyes. This filling of the canal of Schlemm with blood is more difficult to produce with the modern gonioscopic contact lenses, such as the Goldmann or Thorpe-Allen lenses. Blood is also seen in the canal of Schlemm in iridocyclitis and other inflammations of the anterior segment of the eye.

In my two cases, venous congestion in the ciliary plexus caused increased venous pressure in the intrascleral and deep scleral plexuses into which aqueous is drained from the canal of Schlemm via the external collector channels. This increased venous pressure near the external wall of the canal of Schlemm forced blood into the canal and into the trabecular spaces. In Case 2, the blood broke through the trabecular spaces and trickled to the bottom of the anterior chamber, where it gave rise to the hyphema. The presence of blood and decomposed blood products in the canal of Schlemm and in the trabecular spaces may be responsible for permanent changes in the outflow channels. These changes could possibly be responsible for the maintenance of high tensions in the eye and lack of response to treatment. The possibility of a vascular thrombus filling the canal of Schlemm and blocking outflow of aqueous should also be kept in mind.

Treatment of hemorrhagic glaucoma with retrobulbar injections of procaine and priscoline was first advocated by Stern[1] who, in 1954, reported good results in two similar cases. Stern explained the relief of pain after the retrobulbar injection of priscoline by the vasodilator action of the drug on the spastically constricted vessels of the ciliary body. This, according to him, relieves the pain caused by ciliary congestion but does not have any effect on the elevated tension. The addition of procaine may well be responsible for the initial and prompt relief of pain by paralyzing the ciliary ganglion. Topical steroid therapy clears the iritis and seems to be an important factor in the treatment. It is difficult to explain why the high intraocular pressure is not lowered by large doses of Diamox. Becker[2] has also observed that this type of glaucoma reacts poorly to Diamox. It is possible that, in view of the vascular stasis in the eye, Diamox does not even reach the ciliary processes, where it could exert its specific action of inhibition on the formation of aqueous.

The problem of surgical treatment of hemorrhagic glaucoma deserves more attention than it has received. The reason for the defeatist attitude toward surgical intervention in hemorrhagic glaucoma undoubtedly comes from the fact that many eyes have been lost from severe postoperative bleeding after such surgery. This is especially true in those eyes in which there is an old vascular membrane lining the periphery of the iris and the angle together with a long-standing hyphema. But not all cases of hemorrhagic glaucoma present such a clinical picture. After the iritis was cleared, my two cases had white noncongested eyes without vascular membranes. Therefore, in these two cases, it may well be that a cyclodialysis might have reduced the high tension without

serious postoperative complications.

In other instances, I have noted cases of hemorrhagic glaucoma with a thrombosis of the central retinal vein in which, however, the acute congestive attack of glaucoma was due to a coincidental closure of the angle in a narrow-angle eye. In such cases the hemorrhagic glaucoma may mistakenly be attributed to the central retinal vein thrombosis and surgery may, therefore, be unwisely avoided. As a matter of fact, an iridectomy will control the tension in such cases and will result in a quiet eye despite the presence of hyphema.

In another case I have noted an acute attack of congestive glaucoma which resulted from a swollen hypermature lens. The eye presented the picture of hemorrhagic glaucoma in the sense that there was hyphema and neovascularization of the iris. I found that the glaucoma was due to a phacogenic iridocyclitis and the hyphema was due to bleeding from congested and sclerotic iris vessels in an elderly patient. In spite of the hyphema and the iridocyclitis, a successful intracapsular lens extraction was performed and the eye quieted down shortly after the operation. There was no postoperative bleeding.

These examples show that in some cases of hemorrhagic glaucoma surgical intervention is indicated.

Summary and Conclusions

1. Two cases of hemorrhagic glaucoma are presented in which the acute congestive attack occurred in eyes with wide angles.

2. In both cases the attack was triggered by an acute iritis.

3. In Case 1, there was a pre-existing rubeosis of the iris; in Case 2, there was only mild diabetic retinopathy.

4. In both cases the canal of Schlemm was filled with blood. The significance of the engorgement of the canal with blood is discussed.

5. Evidence was presented that corneal edema and pain may not be due to the high tension alone but rather to the iridocyclitis. Pain and corneal edema cleared when the iritis cleared and did not return in spite of persistent high tension.

6. A retrobulbar injection of one cc. each of procaine (two percent) and priscoline followed by topical steroid therapy was instrumental in obtaining a white painless eye.

7. Indications for surgery in hemorrhagic glaucoma are discussed.

885 West End Avenue (25).

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LENS EXTRACTION BY TRACTION

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A method of cataract extraction, which allows traction to be used to good advantage throughout the procedure, is described in this paper. We find that the zonular attachments are freed relatively easily over the first 180 degrees by either the tumbling or the sliding methods. If iatrogenic complications are to develop, they are more likely in the freeing of the latter 180 degrees of the zonule. It is during this part of the procedure that pressure becomes less valuable and of greater danger. The globe will be soft at this time because (1) the lens offers diminished support to it and (2) the operator has used modern akinesia. For this reason traction and pressure in the usual manner are also relatively less effective.

The maneuver which allows the operator

to preserve the advantages of traction in the last stages of the delivery of the lens involves the placement of a traction suture at the limbus in the 6-o'clock position before the corneoscleral section is made. The cataract is delivered by the sliding method. When the upper 180 degrees of the lens have been freed of their zonular attachment, traction and rotation are maintained.

If the remaining zonule is fragile, nothing more may need to be done, as the lens is delivered without further ado. Feeble help is sometimes obtained with the outside hand holding a blunt instrument. This instrument appears to be able to effect a "grasp" on the tissues of the limbus and then it may be used as an instrument for traction.

When these maneuvers appear to lack sufficient mechanical advantage, the traction suture is grasped with the plane forceps in the outside hand, and the lens delivered with this point of counter traction acting against the lens grasped by forceps or erisophake.

Method of akinesia

Analgesia is usually obtained during the procedure by means of Demerol and Phenergan given in divided doses before and during surgery. The motor action of the facial nerve is blocked by the usual methods.

Retrobulbar injection is made with a solution prepared in the following manner: 150 turbidity reducing units of hyaluronidase is dissolved in one-cc. of sterile distilled water. One half of this solution is drawn up into a five-cc. syringe. A 100 mg. vial of spinal procaine is dissolved in two-cc. of sterile distilled water. The 0.5-cc. of Hydase and the two-cc. of procaine are combined, giving a freshly prepared solution containing about 60 mg. procaine and 45 units Hydase in the retrobulbar injection. This is evidently a hypertonic solution.

Bulbar pressure is then maintained for five minutes as recommended by Chandler.

A single lid suture is placed in each lid, the assistant retracting the lids by means of these sutures. A straight hemostat is placed, crushing the tissues of the lateral canthus, and a canthotomy is done as a matter of routine.

Placement of the traction suture

A 4-0 black-silk suture is placed in the globe about one-mm. behind the limbus at the 6-o'clock position. A firm bite is obtained in the superficial layers of the sclera. At least two-mm. of firm scleral fibers are included in this bite. A small peritomy will sometimes be necessary to assure accuracy in the passage of the needle. The thread is then brought through so that, when doubled, it reaches a length of 16 to 18 cm. A double knot is placed in this suture at a distance of one-cm. from its point of insertion. The end of the suture is then tagged with a serrafin. A superior rectus suture of blue nylon is then placed and tagged with a serrafin.

The section

A fornix-based flap is then made. A Graefe section is made and enlarged with scissors. Bleeders are then coagulated by application of Hildreth's cautery. I believe that this instrument is fully and safely effective if used in the following manner: the bleeders are located under $\times 2$ magnification and the cautery applied no closer than two or preferably three-mm. from the wound edge. When used in this manner, hyphema is rare. Application of the cautery to the wound edge invites hyphema and filtration.

The iridectomy

Two perfectly round iridectomies are easily made at the 11- and 1-o'clock positions, using Chandler's excellent instruments to grasp the iris. This little toothless forceps allows cosmetically attractive peripheral iridectomies to be done without risk of injury to the lens capsule. When the iris has a deep valley proximal to the collarette it is easy to injure the lens capsule if the iris is grasped in this portion with a toothed forceps.

Extraction of the lens

Pressure over the lower cornea with a strabismus hook causes the upper portion of the

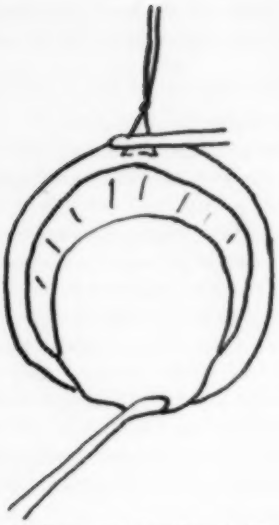

Fig. 1 (Sullivan and Anneberg). The traction suture is held snugly by the outside instrument. The lens is being pulled in the opposite direction by the instrument which grasps it. The zonular fibers in the region of the suture may be freed in this way.

lens to tilt in the direction of the wound. A forceps of Arruga or Kirby design grasps the capsule anterior to the equator at the 12-o'clock position. The technique described in much detail by Kirby[2] is used to initiate separation of the zonule, if such separation is not spontaneous. The following maneuvers are used further to separate the zonule: downward and forward traction, and lateral rotary traction in the plane of the iris. The lens may be lifted forward from the patellar fossa 30 degrees or more. The maneuver next to be described is preferred, however, to extreme forward tilting of the lens.

Application of traction maneuver

Continuation of traction rotation, gently lifting of the lens from the patellar fossa, and rotating it in the elevated position—these procedures may suffice to deliver the lens of the zonule is not too tight. Since direct separation is made less safe in the zone where the dome of the cornea is intact, the alternative is sought in the use of the limbal traction suture.

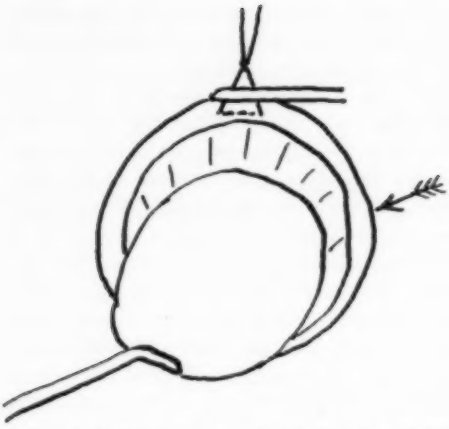

Fig. 2 (Sullivan and Anneberg). Rotation of the lens with continued traction. The zonular attachments in the region of the arrow will be loosened by traction in this direction.

The grasp on the lens is maintained by the erisophake or the forceps, and the outside hand engages the traction suture very close to the globe with a plane forceps. The assistant maintains elevation of the corneal flap. Counter traction is maintained on this suture while traction and rotation of the lens

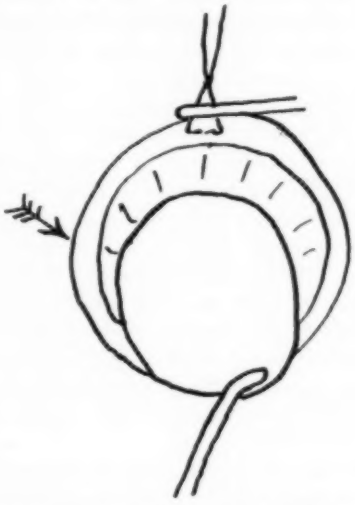

Fig. 3 (Sullivan and Anneberg). Rotation of the lens with continued traction in the opposite direction. The zonular attachments in the region of the arrow should be loosened in this way.

proceed in the plane of the iris. By this means the operator will feel the gentle separation of the lower 180 degree of zonule between the two tissue-engaging areas.

There is no backward pressure, the traction suture actually pulls the tissue of the lower limbus in a direction away from the wound. The effect is the opposite of pressure on the lower pole of the lens.

The vitreous face appears to be protected because (1) the lines of force are parallel to it, (2) the lens is parallel to the vitreous face and making no pressure on it, (3) all instrumentation is remote from the vitreous face.

As soon as the lens is free from zonular attachments the grasp on the counter-traction suture is released and the forceps is free to draw at the corneal-scleral sutures. A similar suture was described by Kirby[3] but was for the tumbling operation. We claim originality in that (1) the suture is used in the intracapsular extraction of cataract by the sliding method, and (2) it is used to facilitate the separation of that portion of the zonule inaccessible to direct maneuvers.

Comment

The suture for counter traction has been placed prior to cataract extraction in 12 cases. It has proved useful in six cases. The effectiveness of the counter traction suture is attested by the ease with which zonular separation occurs. No vitreous was lost during the execution of the maneuver. No capsular rupture occurred.

Complications occurred in two of the cases in which the traction suture was in place but not used. In one case, following complete iridectomy, a vitreous bleb appeared. The lens was looped and the safety sutures drawn up, with minimal vitreous loss. This lens measured six-mm. in diameter and was plano-convex in shape, being flat on the posterior surface. The pupil did not dilate well in the office or prior to surgery. In the second case, under general anesthesia for extraction of a mongolian cataract, a pinhead-sized vitreous bleb appeared in one of the peripheral iridectomies immediately upon separation of the zonule above. The lens was delivered by traction and sliding rotational maneuvers without recourse to the suture. Vitreous loss was again minimal as the safety sutures were drawn up.

It is considered that these complications bore no relation to the placement of the traction suture at the inferior limbus.

502 North Court.

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PERFORATING WOUNDS OF THE CILIARY BODY

A study of 45 cases

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Injuries of the ciliary body are of particular interest due to the marked inflammatory reaction which they generally produce as well as the possibility of causing sympathetic ophthalmia in the fellow eye.[1] Although many studies of perforating wounds of the globe have appeared in the literature during the past 30 years, very few have made special reference to wounds of this region.

The present paper deals with a series of 45 consecutive cases of this type of trauma which were seen in the years 1953 to 1957. All of these, except two, were due to industrial accidents. An analysis on the basis of associated findings as well as retention of the globe is as follows:

Associated intraocular foreign body	19	(42%)
Associated corneal, iris, or lens injury	15	(33%)
Immediate enucleation	5	(11%)
Enucleation after attempted repair	8	(8%)
Retained the injured eye	31	(66%)
Sympathetic ophthalmia		None

Initial hemorrhage was usually considerable. All intraocular foreign bodies over four mm. which passed through the ciliary body were accompanied initially by poor red reflex or no red reflex in our series. Persistent vitreous hemorrhage in five cases accounted for reduced visual acuity and in two cases was thought to be responsible for retinitis proliferans and detachment. In large wounds hyphema was the rule. Those cases with massive hyphema were noted to have disorganized globes, on section.

Infection did not seem to play a prominent part in results with the exception of five cases which had an early endophthalmitis. All had wounds from dirty or greasy material.

Almost without exception all cases had an iridocyclitis of varying severity necessitating a somewhat longer hospitalization time than usual. Three were of such severity that the eye later became phthisical. The longest hospital time was 27 days and the shortest four days with an average of 15 days, in spite of the fact that a number had enucleation within a short time.

The pathologic findings on enucleated eyes were as follows:

Endophthalmitis	6
Disorganized globe	4
Chronic iridocyclitis	2
Phthisis bulbi	2

Most eyes were observed to be very soft to tactile examination excluding those which had large gaping wounds or which had obviously lost intraocular contents. This is in accord with the findings of Weekers[2] who repeating experiments of others found a fall of intraocular pressure of about 15 mm. Hg (Schiøtz) after penetrating the ciliary body of rabbits with fine needles. He attributed this to a reflex vasoconstrictive effect. However, two cases developed secondary glaucoma, one of which was controlled with Diamox and miotics. Although cyclodialysis is frequently reported in these types of injuries it was not noted in this series.

Two factors had a considerable influence on the eventual outcome; the size of the associated foreign body when one was present and the size and nature of the laceration of the ciliary body. Large foreign bodies, those over five mm. in any diameter, passing through the ciliary body were accompanied with blindness with one exception. The exception was a young man with an intraocular foreign body 5.0 by 7.0 mm. who had a final corrected vision of 20/60 three years after injury. His vitreous is still hazy and retinitis proliferans may supervene. One case had a retained intraocular foreign body in the ciliary body 1.0 by 1.0 mm. which has remained in situ for over three years without the eye showing the slightest reaction. All investigations pointed to this particle being tool steel. Operations were unsuccessful on two occasions.

Scheie,[3] in 1952, advised that prolapses should be excised or cauterized before suturing. He also noted the poor results obtained when the wound is large or jagged and counseled prompt enucleation when light perception was poor. Snell,[4] in 1945, showed a loose relationship of length of laceration to visual outcome and this was definitely our experience. Those cases amputated did as poorly as those replaced in our group.

Treatment

All patients received typhoid vaccine in amounts varying from 10 to 25 million units in 250 cc. saline by intravenous drip. This was repeated or increased every third or fourth day depending on the severity and duration of the iridocyclitis. Almost all patients received ACTH in doses varying from 20 to 40 units daily. Three patients had serious complications resulting from ACTH. One developed a symptomatic peptic ulcer proven by X-ray examination which took several

months to heal. One developed a psychosis which necessitated treatment in a state hospital for eight months. We were unable to evalute completely the role of ACTH in the latter case. One patient developed diabetes which took some months to control. Local treatment consisted of atropine, steroids, 10-percent Neosynephrine in some cases. A few received local antibiotics as well. No systemic antibiotics were given.

Immediate surgical repair of the sclera was done on all cases in which there was any possibility of loss of contents. No foreign bodies were removed through the wound of entry in the ciliary body except for those lodged in it. These were removed through the original wound or approached through the angle. Most all posterior foreign bodies were removed through the ora serrata.

Results

Eleven patients (24 percent) achieved a final or corrected vision of 20/30 or better in the injured eye. All of these were cases in which the ciliary-body injury was minute, usually from a small foreign body passing through or lodged in the ciliary body.

Visual results

20/20 to 20/30	11
20/30 to 20/200	7*
20/200 to hand movements	1
Hand movements to blindness	12
Enucleations	14

Causes for reduction in visual acuity

Persistent vitreous hemorrhage	5
Irregular astigmatism	4
Cataract	3
Retinitis proliferans	2
Retinal detachment	2
Secondary glaucoma	2
Macular changes	1
Unknown	1

Summary

Observation and results in 45 consecutive cases of perforating wounds of the ciliary body seen over a four-year period are tabulated. The guarded prognosis in these injuries is noted, with less than 25 percent achieving vision of 20/30 or better. It is suggested that an internist also see the patients who are to be on ACTH for any protracted period.

6 North Michigan Avenue (2).

* One patient died of carcinoma of the stomach while under observation but achieved a final vision of 20/40 on his last visit.

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NEUROBLASTOMA*

A case report

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This paper records a rare case of neuroblastoma in the older age group.

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Case report

A 62-year-old Hindu man, attended the hospital in December, 1957, with the complaint of proptosis of both the eyes for the last one and a half years. The left eye was involved first. There was nothing noteworthy in the past or the family history.

On examination the patient was fairly nourished but anemic. Local examination revealed a greater proptosis of the left eye. A tumor was bulging under the skin of the lids with a few prominent veins overlying it. On palpation the tumor was rubbery hard in consistency embedding the eyeball completely from all sides. Although the overlying skin and

Fig. 1 (Mathur). Appearance of patient.

orbital bones were free, the tumor extended as far back into the orbit as it could be palpated. The lids were slightly edematous and the conjunctiva chemosed, due to exposure. The cornea was completely covered by the upper lid and there was no exposure keratitis. Pupils were semidilated and sluggish in reaction.

There was lenticular sclerosis due to which visual acuity was reduced to finger counting at one meter and the fundus could not be seen. Ocular movements were totally restricted. Lacrimal glands were seen bulging out of the lacrimal fossa. The left preauricular and submandibular lymph glands were enlarged, the left side of soft palate was depressed, and the nares were blocked. No tumor was palpable in the abdomen.

X-ray study. The long bones normal. The skull showed a soft tissue shadow in both the orbits, maxillary antra, and nasal cavity. The chest showed thickened right interlobular pleura due to old pleurisy and a calcification shadow of the arch of the aorta which is not unusual at this age, due to atherosclerosis.

Blood. Hemoglobin 4.0 percent. RBC, 25,000,000 per cmm. WBC, 12,505 per cmm. Polymorphs, 72 percent, lymphocytes, 28 percent, no immature cells. ESR, 110 mm. after one hour Westergren. Bleeding time, 5.0 minutes, coagulation time, 4.5 minutes. Kahn test negative.

Tissue was removed for biopsy from just under the skin of the lower lid of the left eye. It was rubbery hard in consistency and the raw area bled profusely. Microscopically, due to presence of rosettes and fibrillae and characteristic appearance of the cells, it was reported to be neuroblastoma.

The patient was given the prognosis and was advised to take radiation therapy which he refused and left the hospital to die in peace among his kith and kin, rather than to submit to treatment. He was followed for six months at monthly intervals, when he died. He did not develop any swelling in the abdomen or metastases anywhere in the body, only the local spread of the tumor. Autopsy could not be performed.

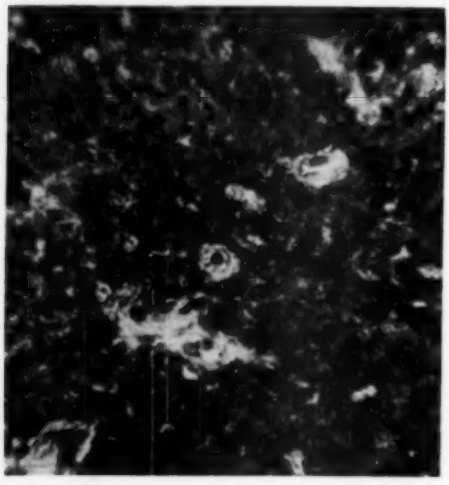

Fig. 2 (Mathur). Photomicrograph, showing a rosette with cells arranged in a concentric manner. In the center is an eosinophilic fibrillar stroma. Cells are round or oval, with moderately stained basophilic nuclei almost completely filling the cells. These masses of round cells are invaded at places by eosinophilic fibrous tissue bands cut in various planes.

Discussion

Neuroblastoma is a highly malignant tumor arising from sympathetic neuroblasts from the medullary plate, thus it may originate from sympathetic cells situated anywhere in the body. The tumor usually is primary in the adrenal medulla. In this case it seemed to be primary within the orbits.

The age group is usually between nine months to nine years and in stillborn babies, very rarely in adults. Halpert, et al.[1] (1953), reported a patient, aged 53 years, in whom tumor appeared to arise from the anlage of the adrenal medulla just outside the adrenals. The present case was in an old man, 62 years of age. Patients usually die within months, while this patient survived for at least two years. It may be that the malignancy of the tumor is reduced in advanced age. This is further stressed because the tumor, which usually spreads to the sturnum, ribs, long bones, and liver, did not spread much in this case except to the regional glands and surrounding air sinuses.

Wright,[2] in 1910, was first to show that the presence of fibrillas and rosettes was characteristic. The tumor consists of small cells with hyperchromatic nuclei which are

polymorphous, contain little cytoplasm, and lay in dense newly formed connective tissue. This description was duplicated in the present case so that the diagnosis of neuroblastoma was beyond dispute.

SUMMARY

An unusual case of neuroblastoma is described. It lasted for two years and did not spread except to the regional lymph glands and air sinuses. No tumor could be discovered in the abdomen. It is suggested that the tumor might have originated from the sympathetic tissue within the orbits.

Victoria Hospital.

I wish to thank Dr. P. N. Wahi, principal and professor of pathology, S. N. Medical College, Agra, for his biopsy report. I thank P. M. and H. O. Bharatpur for permission to publish this report and Dr. M. N. Saxena for his keen interest during investigation and discussion of the case.

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SIGNIFICANCE OF PHTHISIS BULBI IN RETINOBLASTOMA*

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So prevalent is the concept that retinoblastoma does not occur in a phthisical or smaller than normal eye, that it has become a general teaching principle in the differential diagnosis of this disease (Danielson,[1] Manschot,[2] Hughes[3]). Mention of the occurrence of retinoblastoma in normal-sized eyes with normal anterior-chamber depth by Reese[4] in a short two-sentence paragraph seems to be quoted in the literature to the neglect of his more emphatic warning that in a young child a phthisical eye that cannot be accounted for by some known pathologic process may harbor a retinoblastoma.

Retinoblastoma is said to be considerably more rare in Negroes than in whites (Duke-Elder[5]), although no specific statistics are available. The tumor is said to occur bilaterally in between 25 and 33 percent of cases (Reese,[4] Duke-Elder,[5] Carbajal,[6] Herm,[7] Jain[8]). The mortality rate in bilateral cases is about 50 percent (Reese,[4] Carbajal[6]). Unilateral cases have been said to have an equal mortality rate by Reese,[4] Herm, et al.,[7] and others, but in a recent study by Carbajal[6] in which there were 22 bilateral cases and 50 unilateral cases the mortality rate was 54 percent for the bilateral group and 20 percent for the unilateral group.

CASE REPORT

Cook County Hospital Case 58-03911. W. W., a 13-month-old, full-term Negro female, was first seen in the eye clinic on December 27, 1957, referred from the pediatric out-patient service, because of a whitish reflex at the left pupil, thought to be a cataract. A history of having had a respiratory infection at about six or seven months of age was obtained. The mother thought that prior to that illness the baby could see because it would reach for objects. She further stated that she did not think that the baby had been able to see for at least two months prior to her first visit to the eye clinic.

Examination of the right eye revealed a definitely small soft eye with deep scleral injection. The baby was noted to rub the eye frequently. The cornea was clear. The anterior chamber was shallow. There was no pupillary response to light, and the eye did not follow a light stimulus. The pupil was irregular and a number of posterior synechias were noted. There was a gray pupillary membrane involving an opaque lens. The posterior segment could not be visualized. Attempts at transcleral illumination failed to reveal any perceptible light at the pupil.

Examination of the left eye revealed a normal-sized eye which was definitely hard (Schiøtz tension at surgery was 42 mm. Hg). There was no pupillary response to light nor was there any following of a light stimulus. The conjunctiva was not remarkable. The cornea was slightly steamy but examination was easily accomplished after clearing with glycerine. The anterior chamber was very shallow. The pupillary margin was bound by a number of posterior synechias. A white reflex appeared at the

* From the Department of Ophthalmology, Cook County Hospital. Read before the Chicago Ophthalmological Society, May, 1958.

pupil and examination revealed it to be a retrolental membrane or mass, having a nodular or multiple plaquelike appearance. The posterior segment beyond this could not be seen. The eye failed to show perceptible light at the pupil to transcleral illumination.

Roentgen films of the skull in posterior and lateral views revealed scattered calcifications in both orbits corresponding to the position of the globes.

Because of the dense retrolental mass or membrane, the left eye was thought to harbor a malignant growth and was enucleated on January 22, 1958. After obtaining a positive diagnosis of retinoblastoma from paraffin sections of the left eye, the right eye was enucleated on February 3, 1958.

*Pathology.** (Northwestern University, Department of Ophthalmology, Laboratory #58-7, left eye.) Cross section through the optic nerve (paraffin): No evidence of tumor cells.

Paraffin section. The cavum oculi is filled with a tumor mass consisting of necrotic cells. However, there are quite a number of areas consisting of viable tumor cells. The cytologic elements consist of darkly staining round cells with very little cytoplasm. There are some areas of pseudorosettes. The tumor, in some areas, has invaded the choroid. There are some areas of calcification.

Diagnosis. Retinoblastoma.

(Northwestern University, Department of Ophthalmology, Laboratory #58-14, right eye.) Cross section through the optic nerve (paraffin). No evidence of tumor cells.

Paraffin section. The entire cavum oculi is filled with a tumor mass consisting of round dark-bluish cells with scanty cytoplasm. There are numerous irregular mitotic figures. There are quite a number of true rosettes. There is minimal infiltration of the choroid by tumor cells. There are necrotic areas. The tumor was heavily calcified and required decalcification prior to section.

Diagnosis. Retinoblastoma.

Discussion

This patient was seen by six residents and an equal number of members of the visiting staff of the Department of Ophthalmology. Although retinoblastoma was considered, this diagnosis was held in doubt because of the phthisical right eye. Considered among the other diagnoses were hemangioma, retrolental fibroplasia, and bilateral endophthalmitis.

Steward, et al.,[9] have reported a case of spontaneous regression of retinoblastoma and gathered some 15 other cases from the literature. Among these 16 cases four were mentioned as having phthisical eyes. Sovik[10] reported bilateral retinoblastoma in six siblings; three had phthisis bulbi of one eye.

Jain,[8] reporting 76 cases of retinoblastoma, noted two instances in which the eye shrank after degeneration, one after radiation, and two cases after the globe perforated. Eliminating the case of phthisis following radiation, this would make an incidence of 3.94 percent of phthisical eyes.

Carbajal[6] presented 72 cases of proved retinoblastoma with one case of phthisis bulbi and another said to be "approaching the state." Considering this as two cases the incidence of phthisical eyes would be 2.77 percent.

From these data it would appear that a small but significant number of eyes bearing retinoblastoma become phthisical, however, the majority of reports are not analyzed specifically for this and in the literature this fact is usually casually buried in case reports.

Generally, these eyes have certain common characteristics. They are usually described as bearing tumors of large size which showed much necrosis or had undergone complete regression. The mechanism for the phthisis has been suggested by Duke-Elder[5] and Reese.[4] With rapid increase in the size of the tumor, the blood supply cannot support the high metabolic activity of the mass and it undergoes necrosis, resulting in a reactive degeneration of the ocular tissues to the necrotic tumor tissue.

It has been stressed by Duke-Elder,[5] Reese,[4] Sovik,[10] and Steward[9] that such phthisical eyes should be enucleated and not retained in hope that complete spontaneous regression will occur.

Summary

1. The concept that retinoblastoma is not present in phthisical eyes seems prevalent in the literature.

* The pathologic study was done by Stefan Van Wien, M.D., and Burton M. Krimmer, M.D., Department of Ophthalmology, Northwestern University Medical School, Chicago.

2. A case of retinoblastoma occurring in a phthisical eye is presented.

3. Because the eye was phthisical there was some doubt about the preoperative diagnosis of retinoblastoma.

4. A number of cases of retinoblastoma in phthisical eyes have been collected from the literature. It is concluded that there are a small but significant number of cases of phthisis bulbi with retinoblastoma.

5. The mechanism of phthisis with retinoblastoma is a reactive degeneration of ocular tissues to the necrosis of a regressing tumor.

6. All phthisical eyes that occur in infants or young children should be enucleated, if the shrinkage of the globe cannot be definitely attributed to some known benign pathologic process.

8954 North Keeler Avenue.

ADDENDUM

Five months after enucleation of the left eye, the implant in the left orbit was removed because it was displaced superiorly behind the upper lid. Biopsy of the orbital tissue proved positive for tumor cells and X-radiation was begun to the orbit. One month later the child suddenly died. A marked anemia was noted a few days prior to death. Unfortunately a post-mortem examination was not obtained. The final outcome of this case emphasizes the necessity of early diagnosis and treatment of this highly malignant tumor.

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THERAPY OF SENILE MACULAR DEGENERATION

EDWARD O. BIERMAN, M.D.
Santa Monica, California

INTRODUCTION

Soon after Rome* reported the treatment of macular degeneration by intravenous injections of aqueous heparin (100 mg.) twice weekly for five to 10 weeks, evaluation of the procedure was begun. The techniques I employed were, as closely as possible, those used by Rome. In addition, a series of patients treated with vitamins and other supportive therapy was used as a control group. Rome had reported no control studies.

DATA

Thirteen patients were treated with heparin; 10 were treated supportively and followed as controls. Of the 13 treated with heparin, one patient showed great improvement, two appreciable improvement, eight no appreciable change; and two patients became worse. In addition, one patient suffered a "heart attack" during therapy, and another suffered subcutaneous hemorrhages in both forearms.

One must remember that in this age group anything might happen, and it is impossible to place the responsibility on the medication. However, if lipoid deposits are acting as a protection to the vessel wall and only inadvertently as an obstructing agent, their removal might cause hemorrhages. Lipoid deposits may provide nature's repairs.

* A case presented to the Los Angeles County Medical Association, May, 1956.

TABLE 1

SENILE MACULAR DEGENERATION TREATED WITH INTRAVENOUS HEPARIN THERAPY

Case	Sex	Age (yr.)	No. of Injections	Vision		Pathology*	Results
				Before	After		
1	M	84	10	O.D., fingers 6 ft. O.S., 20/60−	20/400 20/50+	H & D D	Improvement
2	M	81	4 (stopped because heart attack; survived)	O.D., 20/80 varies O.S., no. L.P., glaucoma	20/40 varies widely	D & H	No change
3	M	86	10	O.D., fingers 2 ft. O.S., fingers 2 ft.	fingers 2 ft. fingers 2 ft.	D & H D & H	No change
4	M	77	20	O.D., fingers 2 ft. O.S., fingers 2 ft.	20/200 20/50	D & H D & H	Improved
5	F	69	8 stopped because of subcutaneous H	O.D., 20/400 O.S., 20/25	20/400 20/20	D & H D	No change
6	F	70	10	O.D., 20/300 O.S., 20/70−	20/200 20/80	D & H D & H	No change
7	M	76	10	O.D., 20/400 O.S., 20/400	20/400 20/400	D & H D & H	No Improvement
8	F	68	20+	O.D., 20/300 O.S., fingers 4 ft.	20/200 20/200	H D & H	Improved
9	F	74	11	O.D., 20/30 O.S., 20/25	20/30− 20/30+	D D	No change
10	F	75	10	O.D., 20/80 O.S., 20/60	20/400 20/400	D D	Worse
11	M	80+	10	O.D., fingers 4 ft. O.S., fingers 4 ft.	20/400 fingers 4 ft.	D D	No change
12	M	80	10	O.D., 20/25− O.S., 20/60−	20/20−2 20/70	D D	No change
13	M	74	5 (stopped at patient's request)	O.D., 20/300 O.S., 20/20	20/300 20/40+	D D	Worse

Totals: One much better, two some better, eight no change, two worse.
* D—degeneration; H—hemorrhage.

In the control series, one patient was much improved, three were better, questionable change was present in five, and two patients were worse. No undesirable side-effects occurred during the period of testing. The results as shown in Tables 1 and 2.

Discussion

The tables indicate a great similarity in the heparin and control series. Heparin seemed no better than usual supportive measures and was more expensive and troublesome. No serious hemorrhages occurred from its use and its side-effects were minimal, as reported previously.

These cases revealed how difficult adequate evaluation of such cases can be. The progress of the disease is not steadily downward. It resembles diseases of the heart in that some days and months are excellent, while others are worse. The general trend is downward over a long period of time but the periods of temporary improvement confuse evaluation of therapy.

Degenerative changes are usually slowly disabling but a hemorrhagic phase is immediately apparent at times. In our series, the absorption of hemorrhage with or without heparin is the most important feature of the condition. With absorption improvement generally occurs. However, in one case in which only degeneration was present, improvement occurred after supportive treatment. Similar improvement was seen in one patient without hemorrhages in the heparin series.

Suggested treatment

The psychologic importance of any treatment is great. Experience has proven that

TABLE 2

CONTROL SERIES SUPPORTIVE THERAPY

Case	Sex	Age (yr.)	Vision		Pathology*	Results
			Before	After		
1	F	74	O.D., 20/25 O.S., fingers 2 ft.	20/25 20/50−	D D	Greatly improved
2	F	65	O.D., 20/40 O.S., 20/60	20/30 20/40+	D D	No change
3	F	74	O.D., 20/200 O.S., 20/20	20/25+ 20/70−2	D & H D	No change
4	F	75	O.D., 20/30 O.S., 20/100	20/25 20/70−2	D D & H	No change
5	F	83	O.D., 20/50− O.S., 20/300	20/50+2 20/70+	D D & H	Fluctuates widely; followed 3 years
6	F	74	O.D., fingers 1 ft. O.S., 20/30	20/100 20/20	D & H D	Improved
7	M	75	O.D., 20/50 O.S., 20/50	20/25− 20/30	D D	Improved
8	F	75	O.D., 20/40− O.S., 20/20	20/60 20/25	D D	3 years, slightly worse
9	F	76	O.D., 20/400− O.S., 20/400−	20/400− 20/400−	D D	No change
10	M	66	O.D., 20/200 O.S., fingers 2 ft.	20/400 20/70	D & H D	Better in one eye, worse in the other

Totals: Three better, five no change, two worse.
* D—degeneration; H—hemorrhage.

the value of assurance to a patient possibly exceeds the value of the medication. The patient is told:

1. Blindness, as it is interpreted by the lay person, will not occur. Loss of fine vision may but the patient will always be able to fend for himself.

2. The condition can improve, although the patient may have a remission. This happy thought means much to many who have been told they will go blind and that nothing can be done.

3. The disease progresses very slowly, barring accident (hemorrhage). Most patients do not feel they have long to live anyway and feel their eyes will outlive them when told this.

I use supportive treatment first. Heparin is used in desperation to improve a patient's attitude, as well as his ocular condition for his sake and for that of his family.

SUMMARY

1. Supportive treatment with vitamins and, occasionally, nicotinic acid proved as effective as heparin intravenously in my cases.

2. Supportive therapy and assurance is the first treatment utilized. Heparin is now used in patients who have "benefited" from it and in especially desperate cases.

3. The psychologic aspects of the condition are of great importance.

A KERATOME OF NEW DESIGN

EDWARD GROM, M.D.
Caracas, Venezuela

Surgeons who make a keratome incision in limbus in cataract operations generally use keratomes of different widths but with a length of approximately 12 mm. The size of such keratomes is troublesome because (1) the instrument does not go deep into the anterior chamber, resulting in a narrow incision which is difficult to widen with scissors; (2) the length of the keratome makes it difficult to avoid wounding the anterior capsule of the lens in the pupillary area.

To avoid these annoyances I have designed a keratome 14 mm. in width and seven mm. in length. Shorter models have too wide an angle, making the introduction somewhat difficult. A keratome of this practical size makes a wide limbal incision which can easily be lengthened with scissors. Because it does not go far into the anterior chamber, an introduction of a few mm. being enough, the lens is not injured.

The model manufactured for me by Grieshaber and shown in Figure 1 completely fills all expected requirements.

Instituo Médico del Este,
Ave. Casanova.

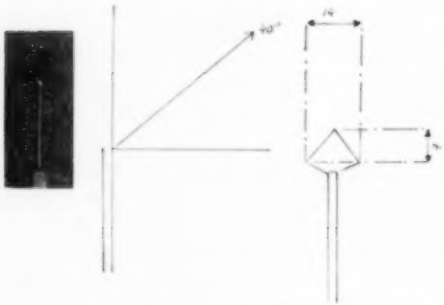

Fig. 1 (Grom). A keratome of new design.

A NEW MULTIPRONGED SCLERAL RETRACTOR*

FOR BROAD TRACTION ON THE EYEBALL DURING SURGERY

CONRAD BERENS, M.D.
New York

Fixation of the eyeball during surgical intervention has been accomplished by traction sutures, fixation forceps, or by a single or bipronged scleral hook.† However, where a firmer, broader hold is required, particularly in lamellar keratoplasty and retinal detachment (scleral resection, scleral buckling, and so forth), these methods of manipulating the eyeball are inadequate. Because rupture of the sclera occurred when traction had been applied over a small area in two cases in which the sclera was thin due to previous diathermy application, a new retractor was devised to overcome this complication by providing a firm grip over a broad surface of the eyeball.

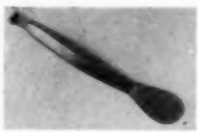

Fig. 1 (Berens). A multipronged scleral retractor which provides traction over a broad surface of the eyeball.

The multipronged scleral retractor‡ (fig. 1) which is 85 mm. in length, is made of stainless steel. The five teeth, set one mm. apart, are two mm. in length and assume a 40-degree angle to the handle of the instrument, which is slightly curved. The flattened corrugated grip on the handle of the retractor permits firm manipulation of the instrument.

708 Park Avenue (21).

* From the Department of Research, New York Association for the Blind and the Department of Ophthalmology, New York University Post-Graduate School of Medicine. This study was aided by a grant from The Ophthalmological Foundation, Inc.

† Berens, C.: Scleral hooks. Am. J. Ophth., **25**: 588 (Apr.), 1942.

‡ Made by Storz Instrument Company, St. Louis, Missouri.

OPHTHALMIC RESEARCH

EDITED BY FRANK W. NEWELL, M.D.

Abstracts of papers presented at the meeting of the Southeastern Section of the Association for Research in Ophthalmology, Inc., Bowman Gray School of Medicine, Winston-Salem, North Carolina, December 5 and 6, 1958

Richard Weaver, *Chairman*
R. Winston Roberts, *Section Secretary*

Test comparing a new with a conventional astigmatic chart. Gerhard A. Brecher, Donald Lewis, and Arthur A. Eastman.

A new astigmatic test chart (Eastman, A. A., and Guth, S. K., Am. J. Optometry, in press) consisting of radiating black lines with white borders on a black and white striped background was compared with a conventional astigmatic test chart of radiating lines. The sensitivity of both charts was measured with a simple optical arrangement by determining the threshold for the test target lines. It was found that the new astigmatic chart was more sensitive than the conventional chart. An end-point defined by "visible" or "not visible" in interpreting charts is considered an additional advantage.

Congenital toxoplasmosis: III. Ocular signs of the disease in state schools for the blind. John R. Fair, M.D., Ophthalmology Division, Department of Surgery, Medical College of Georgia, Eugene Talmadge Memorial Hospital, Augusta, Georgia.

1. A survey of 467 students in state schools for the blind revealed 26 cases of chorioretinitis. Of these, 22 or 84 percent gave positive skin tests for toxoplasmosis compared with 17 percent positive in the remaining 441 students. Twenty mothers were available for serologic study. Fifteen students with positive skin and dye tests had mothers with positive dye tests suggesting congenital toxoplasmosis as the cause for the chorioretinitis in these cases. Three children with chorioretinitis gave negative skin and dye tests, as did their mothers, indicating that some infectious agent other than Toxoplasma may cause congenital chorioretinitis. In two cases, dye tests were negative in the mother of a child with a positive skin and dye test for toxoplasmosis. The significance of this finding is discussed.

2. It is suggested that congenital toxoplasmosis is a public health problem worthy of real concern. Further studies are proposed with the idea of establishing the incidence of the disease.

Present status of corneal preservation. Frederick W. Stocker, M.D., Duke University School of Medicine, and the McPherson Hospital, Durham, North Carolina.

In spite of many attempts to develop a satisfactory method for banking corneal tissue to be used for grafting, the problem is still not solved. Obviously, the method of preservation presently used by most eye-banks in this country; namely, storage in a moist chamber at +4°C., is far from being ideal. Most surgeons refuse to use material which has been preserved in this manner for longer than 48 hours. In order to be able to utilize the available tissue to its best advantage, a long-term storage method would be most desirable. Satisfactory results have been obtained as far as lamellar grafts are concerned with corneas preserved by glycerol-freezing at −79°C. (Rycroft), vacuum dehydration (King) and lyophilization (Payrau).

Unfortunately when corneas preserved by these methods were used for full-thickness grafts, the results were not equally good. It was suspected by me that the delicate endothelium might suffer too much by these methods of preservation to regain its viability. Indeed, my co-workers and I were able to demonstrate that no endothelial growth could be obtained in tissue culture when the tissue had been preserved in that manner. Conversely, positive endothelial cultures were obtained when the corneas were preserved in mineral oil at +4°C. after as long as four weeks and when frozen at −45°C. instead of −79° for as long as 19 weeks. Indeed, European authors feel that it is perfectly safe to use corneas preserved in mineral oil at +4°C. at least up to two weeks. While this constitutes a definite advantage over the storage in moist chamber, a much longer term preservation should be available in order to solve the problem. My coworkers and I presently are engaged in experiments with corneal tissue preserved at −45°C. but so far no definite decision could be reached. The field is still wide open for further research.

Clinical tonography with and without recording galvanometer. R. Winston Roberts, M.D., Department of Ophthalmology, Bowman Gray School of Medicine, and the North Carolina Baptist Hospital, Winston-Salem, North Carolina.

A brief resume of the place of tonographic criteria in the diagnosis and clinical handling of glaucoma is presented briefly. The theory of tonography is briefly outlined and the nature of

the tonogram and its significance are pointed out. Validation of the test by correspondence to other techniques, when performed in a standard fashion with a recording galvanometer and precise technique, is outlined, and the question of whether or not satisfactory tonographic data can be obtained by direct readings from an electronic tonometer without a recording galvanometer is raised.

The material for this study includes 150 cases of which 112 are of proved glaucoma and the rest are in glaucoma suspects. Of these 790 tonograms which were technically good have been chosen for this study. These tonograms had recordings and direct readings from the tonometer, taken at half-minute intervals, done simultaneously in all cases. These data have been analyzed simply and statistically to evaluate the degree of correlation or deviation between the two techniques.

A statistically significant deviation between the two methods is evident, with the high excess of large deviation above the number expected from a normal curve distribution apparently due to fault in the visual technique, with the errors most likely to occur in the first half-minute of the tonography where the effect on the C value will be most marked. These large resulting inaccuracies may lead to completely incorrect classification of the tonogram.

Ophthalmic Miniature

Would you believe me that I left the doctor's office delighted rather than depressed when he had told me that I had a cataract? At last I knew the name of my condition, the nature of the danger which I had to face. This knowledge lifted an enormous weight off my soul. I have since pondered over the question "Should one tell the patient the truth?" It is a moral problem which has tormented me a long time and for which I do not yet know the answer.

Courage, we know, is but the faculty of man to adapt the resources of his mind, his soul and his body to the danger he faces. This adaptation is more or less rapid, according to the circumstances, the temperament and the character, but there is no courage without this process of adaptation. Therefore, imaginary dangers, dangers born out of darkness and night, cause more fear than the real ones. How can one adapt oneself to a danger of which one does not know the form or the name, a danger which has no reality? What can one do against a spectre? Before I knew that I had a cataract I saw vaguely through this failing eye numerous horrible diseases and, at the end, blindness. It was the unknown, the spectre, and I could not adapt myself to a spectre. But the very day I was told "You have a cataract" there was a danger which was classified, known, foreseeable, labeled, real—and at once I could adapt myself to it. I could afford to be courageous.

From *Gare à vos yeux!*, Francisque Sarcey, Paris, 1884.

SOCIETY PROCEEDINGS

Edited by DONALD J. LYLE, M.D.

NEW YORK SOCIETY FOR CLINICAL OPHTHALMOLOGY

May 5, 1958

DR. HARVEY E. THORPE, *President*

HYPOTONY AND ANESTHESIA FOR CATARACT SURGERY

DR. SAMUEL GARTNER said that local anesthesia which completely abolishes all sensation and all motion of the lids and extraocular muscles creates the ideal situation for cataract surgery. This is only partially achieved by the usual techniques of injecting procaine with or without adrenalin for lid block and retrobulbar injection. A more profound degree of anesthesia is achieved by methods that cause a more rapid and extensive diffusion of the anesthetic solution. Hyaluronidase is a very valuable aid. Finger pressure on the closed eye after the injection is an added measure for spreading the anesthetic to reach more nerve fibers. Hyaluronidase and finger pressure induce hypotony, which is an advantage. It requires more pressure to expel the vitreous from a soft eye than a tense one. Hypotony is produced by a number of mechanisms. The paralysis of the extraocular muscle lowers tension about one third. Finger pressure causes the expression of some aqueous, as well as blood, from the eye.

All methods which cause rapid diffusion of anesthesia cause more profound effects but also more rapid absorption, so that the duration of anesthesia is diminished. Methods of increasing the period of profound anesthesia are very desirable. Tetracaine (Pontocaine) has a far longer action than Procaine or Lidocaine and, in the low dilutions that are needed, it is a safe drug. The anesthesia induced by procaine alone is dependable for about 30 minutes to an hour. This is diminished about 30 percent by the addition of hyaluronidase or finger pressure which speeds its diffusion and absorption. The duration of action is lengthened about 10 percent by the addition of adrenalin. Lidocaine has only a slightly longer action than procaine. Tetracaine has at least a 50-percent longer action than procaine. The solution advised for use in cataract surgery is a mixture in these proportions: (1) Lidocaine, one percent; (2) Tetracaine, 0.075 percent; (3) adrenalin, 1:50,000; (4) hyaluronidase, three turbidity units per cc.

Following the retrobulbar, lid-block, and conjunctival injections, pressure is applied for three minutes with a five-second interruption every minute. Tension should then be checked with a tonometer. If it is 12 mm. Hg or less, the operation proceeds. If it is higher, pressure is continued until the desired level of tension is reached. The intraocular pressure serves as a useful guide that can easily be checked. In addition, the routine use of the tonometer before incising the globe will disclose the occasional case in which the tension rises due either to an attack of acute glaucoma on the operating table or a retrobulbar hemorrhage. Dr. Gartner has had three such occurrences in about 200 cases.

The methods advised produce profound anesthesia that is dependable. In addition, hypotony is induced.

Responding to questions, Dr. Gartner said that he had never encountered any difficulty from the routine use of adrenalin in the retrobulbar solution in cases of hypertension. He also said that there was no advantage in using Novocaine in higher concentration than two percent.

CONGENITAL GLAUCOMA

DR. A. EDWARD MAUMENEE reviewed some of the previous theories of the patho-

genesis of congenital glaucoma, and then offered a new theory with evidence that an abnormal insertion of ciliary muscle fibers to the trabecular network in front of the scleral spur causes congenital glaucoma.

In responding to questions Dr. Maumenee gave his opinion that goniotomy was without doubt the best operative procedure for congenital glaucoma. The fact that goniotomy controls tension in congenital glaucoma and cyclodialysis does not supports his theory. He has also observed the same type of abnormal muscle insertion in cases of aniridia.

Tumors of the Iris

Dr. Lorenz E. Zimmerman and Dr. Benjamin Rones of Washington, D.C., presented a progress report on their study of the approximately 250 cases of tumors of the iris on file in the Registry of Ophthalmic Pathology of the Armed Forces Institute of Pathology. A logical clinicopathologic grouping of the cases is:

I. Localized tumors
   A. Which are sufficiently small and discrete to be excised by iridectomy.
   B. Which because of their size, position, or infiltration of adjacent structures cannot be treated by iridectomy.

II. Diffuse tumors
   A. Which have developed as a sequel to IA or IB.
   B. Which present signs and symptoms mainly of glaucoma.
   C. Which produce mainly heterochromia iridis.

To date, Groups IA and II have been studied very thoroughly, while investigation of IB is still in progress.

In Group IA there are 125 tumors which were treated initially by iridectomy. In general, these are small lesions located mostly near the pupil or in the midzonal portion of the iris. Many of the tumors were known to have been present for many years, while others had been discovered only upon recent examination. Patients in this group are mostly young adults, the median age being 40 years. Histologically most of the tumors are characterized by a relatively benign cytology and an infiltrative tendency. Though recurrences may follow incomplete removal by iridectomy, distant metastasis is most unusual. Only three of the 116 patients treated initially by iridectomy who have been followed are believed to have died from metastasis.

Because of their clinical and histopathologic characteristics, small localized tumors of the iris should be treated conservatively. If there has been no definite change in the appearance of the tumor, it is questionable whether those changes which do not involve the peripheral half of the iris need to be treated at all. Those near the iris root should be excised early in order to prevent extension into the ciliary body and anterior chamber angle. Those already in the chamber angle when first examined present problems in management. If the history is one of long duration without change, and if a primary tumor of the ciliary body can be excluded by clinical study, the patient should be followed closely with serial photography and gonioscopy. If treatment is required it probably should be enucleation, though Stallard's suggestion of local excision coupled with corneal transplantation should be given consideration.

Time did not permit Dr. Zimmerman and Dr. Rones to discuss the problem of diffuse melanomas of the iris.

Jesse M. Levitt,
Recording Secretary.

NEW ENGLAND OPHTHALMOLOGICAL SOCIETY

441st meeting, February 19, 1958

DR. VIRGIL G. CASTEN, *Presiding*

CORRECTION OF ENTROPION

DR. FREDERIC B. BREED, Salem, Massachusetts: This is a description of my experiences with a method for the treatment of entropion that is not in general use. In the course of the last seven years about 25 cases of entropion have been treated in the following fashion by me or by residents under my direction. Correction of entropion has resulted in every case. The period of follow-up has been from one to three years.

On the patient's first visit to the office his lid is taped down with Scotch Tape and drops are prescribed for the infection that is usually present. When the patient returns a few days later a drop of pontocaine is instilled into the eye. The skin is painted with tincture of iodine. Two-percent Novocaine is infiltrated subcutaneously and subconjunctivally for the entire aspect of the lower lid, as well as several centimeters lateral to the outer canthus. A lidplate is inserted into the lower cul-de-sac.

A muscle hook is heated cherry-red over an alcohol flame. The muscle hook used is one that is a bit heavier and has a wider shank than those generally used for muscle surgery.

Beginning about three fourths of a centimeter lateral to the outer canthus, a cautery point is made by pressing the heated muscle hook firmly into the lid about four mm. below the line of the lashes. Be certain the lidplate is in position in relation to the hook. Penetration is deep but actual perforation to the plate is not done. If the hook is allowed to cool it will stick. A row of points between five and seven in number are placed, sparing the lower canaliculus. The patient is then sent home without medication or dressing.

In each case it was noted that the entropion no longer existed from the moment of the completion of the operation. A week later some of the patients had enough reaction to cause a temporary ectropion. At the end of a year it was often difficult to tell which lid had been operated on.

CLINICAL COMPARISON OF DICHLORPHENAMIDE WITH DIAMOX

DR. MORRIS HENRY and DR. PEI-FEI LEE, Boston: Dichlorphenamide lowers the ocular tension of both normal and glaucomatous eyes, probably by carbonic anhydrase inhibition in a manner similar to Diamox.

When dichlorphenamide is given orally it has a rapid onset of action, usually within 20 to 40 minutes. It appears to have a longer duration of effect on ocular tension than Diamox.

Approximately one third the dose of dichlorphenamide is needed to produce the same effect as Diamox. Unfortunately, the side-effects were as prominent as those reported when using a comparably effective dosage of Diamox.

We may say then that dichlorphenamide appears to be as effective as Diamox in lowering ocular tension but the side-effects prevent dichlorphenamide from being a superior drug to Diamox, except in cases where the patient cannot take Diamox. In these cases it should be worth while to try dichlorphenamide.

Chlorothiazide (Diuril) was also tested and it did not appear to have any effect on either the ocular tension or the rate of aqueous formation when given orally.

MARCHESANI'S SYNDROME

DR. JOHN C. MCGAVIC, Bryn Mawr, Pennsylvania: In the literature certainly not more than 35 cases of Marchesani's syndrome (brachydactyly with subluxated lenses) are described. The family in this report has had 11 members who have had the syndrome.

The syndrome, first described in 1939, has the following features: Short, stout

build; short fingers and toes; broad chest; good musculature; good subcutaneous fat; reduced mobility of the joints; and brachycephaly. The lenses are small in size and generally round or shaped like the head of a bullet. The subluxation of the lenses occurs later in life and is not present in the first 10 years. There is an index myopia. The patients frequently develop secondary glaucoma. In most reports the inheritance is said to be recessive; in this family the inheritance was dominant.

Mental retardation is said to be a part of this syndrome. The people in this family weren't particularly dull, they seemed to be of average intelligence. There was no known consanguinity in the family.

One of the most interesting things is the appearance of the extracted lens. The zonular membrane strands left on the lens are particularly long. The lens weighs 20 to 25 percent less than the normal lens. The equatorial diameters are 6.75 to 7.0 mm. The sagittal diameters are larger than normal and the lenses tend to project into the pupillary space. This explains the mechanism of the glaucoma on a pupillary block basis.

In this family there were 12 children. The father carried the "bad" gene. Two of the children died in infancy and nothing is known of them. Six of the children had the syndrome. Two of these siblings have two children each and each of these four children have the syndrome. Four great-grandchildren show no evidence to date of having the syndrome.

The syndrome is not manifest unless the pupils are widely dilated. It showed up in this family between the ages of 18 to 20 years. The first sign was myopia, the second was a tremulous iris. The equator of the lens may be visible along with the taut zonule fibers. The next sign was the subluxation of the lens. Secondary glaucoma developed in this family between the ages of 30 to 40 years. An early removal of the lens is advisable; this has been demonstrated by the results in two cases.

This then is a report of 11 cases of Marchesami's syndrome in three generations of one family. Thirteen cases have been previously reported in this country. I think the syndrome is more common than has been reported.

Charles Snyder,
Reporter.

Ophthalmic Miniature

The cause of trachoma is still unknown; neither in the secretions nor in the contents of the follicles have any microbes been demonstrated which can be considered to have causal significance. Even the most modern methods of staining have failed to give any positive result. The latest results of Halberstaedter, Prowozek and Greef regarding the presence of very small granules do not furnish any explanation.

Axenfeld, *Bacteriology of Eye*, 1908, p. 261.

AMERICAN JOURNAL OF OPHTHALMOLOGY

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RETINAL PHOTOGRAPHY*

Skilled ophthalmoscopy yields information not only about local conditions but also about general disorders. But the technique is not easily mastered, and even an experienced ophthalmoscopist finds difficulty in describing his observations. A short period in an ophthalmic department is enough to impress beginners with the differences between reports on a single fundus by a group of experts; and serial observations of a progressive fundus lesion, made in ignorance of previous records, show how open to error the method is. Many progressive retinopathies, such as those associated with hypertension, diabetes, and suspected intracranial tumors, demand more ac-

* Reprinted from *The Lancet,* May 10, 1958, pp. 1008 and 1009.

curate observation than is possible with ophthalmoscopy; and ophthalmologists are therefore turning to photography for objective records.

The Zeiss-Nordenson retinal camera, introduced in 1915, depended for its light source initially on a carbon arc and later on a filament lamp.

An image of this source was formed by optical means at the margin of the dilated pupil and a cone of light entered the eye and illuminated an area of the retina. An aerial image of the illuminated fundus was formed within the instrument by a powerful convex lens as in indirect ophthalmoscopy; and this aerial image was photographed by a built-in camera designed on the principles of a single-lens reflex camera.

The Zeiss-Nordenson camera gave good monochromatic photographs of the retina with exposures of a 10th to a 25th of a second; but such long exposures were liable to cause failures due to blinking and ocular movements; and with color photography (calling for exposures of a half to a fifth of a second) failures were even commoner. Modern developments in lighting have led to successful color photography with fast exposures.[1-4] For this purpose a high-speed electronic discharge flash-lamp has been incorporated in the Zeiss-Nordenson camera, or this camera has been adapted to a xenon-arc lamp. But there is a great demand for entirely new models; and Hansell[5] describes seven which have appeared in the last year or so. Most are based on the Gullstrand ophthalmoscope and Zeiss-Nordenson camera.

Illumination for focusing is usually independent of that for photography. In most, color photographs can be taken by rapid exposure with an electronic flash. One camera compensates not only for refractive errors including astigmatism but also for chromatic aberration in the human eye; 35 mm. film is standard, and most models have standard detachable camera bodies which may be interchanged with other instruments.

Despite all this effort, retinal photographs, even in color, leave much to be desired; and it remains more difficult to recognize a retinal condition from a photograph than from an artist's painting. Admittedly this may be partly because the artist, like the ophthalmologist, has preconceived notions which bias his observations; but the artist can give a satisfying Mercator-like projection of the fundus which resembles the subjective impression on examining the retina. It is difficult to imagine an optical device which could do this—or a suitable method of illumination. In this respect modern retinal cameras are less satisfactory than the original Zeiss-Nordenson camera; for with them the photograph records a smaller area of fundus, and consequently a clinician who needs a record of, say, the whole length of a retinal blood-vessel, must take serial photographs and piece them together.

Some of Leishman's[6] excellent illustrations are composed of 11 separate photographs. A further difficulty for the clinician wishing to use the retinal camera as a research tool concerns magnification. The size of the retinal details depends not on refractive error but on the total refractive power of the eye. For example, the optic disc will be smaller in a photograph of a large emmetropic eye than of a small emmetropic eye. One type of camera makes certain compensations for refractive power; but this modification is intended to give constant size of field—not to compensate accurately for differences in refractive power. No doubt in time these difficulties will be surmounted; but meanwhile we are a long way from achieving the perfect retinal camera.

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NEOMYCIN IN OCULAR INFECTIONS*

Ocular infections continue to contribute substantially to visual disability and blindness, despite the virtual elimination of ophthalmia neonatorum and interstitial keratitis by improved methods of treatment. Most infections are exogenous, and many are associated with injury; bacteriologic cultures would be expected in these circumstances to indicate the appropriate antibiotic or chemotherapeutic agent, but, owing to technical difficulties, culture specimens can be obtained only from the conjunctiva and eyelids, and in more serious corneal and intraocular infections the organism can seldom be detected in this way. Accordingly the nature of the infection must still often be determined largely by clinical deduction, and the efficacy of treatment decided by therapeutic trial or prophylaxis. The value of penicillin used in these ways is evident from the greatly reduced incidence of serious ocular infections (and in particular of hypopyon ulcer) since its general use became possible; but its limitations in respect of gram-negative organisms and penicillin-resistant staphylococci are reflected in the high proportion of serious infections which are now found to be caused by these organisms. Sorsby and Ungar[1] found 19 such infections in a series of 61 inpatients at the Royal Eye Hospital, and they drew attention to the need for some more effective, broader-spectrum antibiotic for use both in such cases and in prophylaxis.

The choice of a broad-spectrum antibiotic presents special problems in ophthalmology owing to the selective action of the blood-aqueous barrier. The comparable levels in blood and aqueous for innocuous sulfonamides are 3.4 mg. per 100 ml. and 0.4 mg. per 100 ml. respectively; with penicillin penetration into the aqueous is better, and still higher levels may be achieved by subconjunctival injection of penicillin with adrenaline.[2,3]

Sorsby[1] now reports trials of several broad-spectrum antibiotics in rabbits infected with penicillin-sensitive and penicillin-resistant staphylococci and with Pseudomonas pyocyanea and proteus. Carbomycin was perforce excluded on account of poor local tolerance; and erythromycin, which also produced some local irritation, gave unpromising results when injected subconjunctivally or given by mouth. The effect of subconjunctival neomycin on penicillin-resistant staphylococci infections was more hopeful. (Additional oral erythromycin produced no material extra improvement.) Ps. pyocyanea and proteus alone among the gram-negative organisms give rise to suitable experimental ocular lesions; and, with the first of these, results comparable with those obtained with polymyxin B and with streptomycin were seen. Intraocular infections with proteus proved impossible to control with any of the antibiotics used, but in a parallel experiment with animals pretreated with neomycin the infection was controlled in most cases, and in a further series of experiments with gram-negative bacteria the less fulminating infections produced by intracorneal injection of proteus were controlled. A final series confirmed that in infections with the common penicillin-sensitive organisms neomycin is as effective as penicillin.

Thus neomycin is no less efficacious against both gram-negative and gram-positive organisms than the other individual antibiotics. Moreover it has greater range; and, being suitable for subconjunctival injection, it may be the antibiotic of choice when the causal organism is unknown. Early clinical experience suggests that the human eye tolerates well the repeated subconjunctival injections of neomycin necessary to maintain therapeutic concentrations, and this antibiotic is clinically effective in severe ocular infections.

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* Reprinted from *The Lancet*, June 28, 1958, p. 1377.

CORRESPONDENCE

La Rocca suture

Editor,
American Journal of Ophthalmology:

In the article "Penetrating keratoplasty" (Am. J. Ophth., October, 1958) I mentioned a suture method described by Lindeman (Klin. Monastbl. f. Augenh., **118**:532, 1951): "A continuous suture is anchored concentrically at several points on the corneal-scleral margin. Loops are left between each point of sufficient length to allow them to be brought almost to the center of the transplant. These loops are firmly connected near the center by a second suture."

I have since found that this identical suture was described by Dr. V. La Rocca of New York 12 years earlier ("The limbal suture," La Rocca, Vito: Tr. Ophth. Soc. U. Kingdom., **59**:[part 2] 739-743, 1939).

The "Lindeman suture" therefore is actually the "La Rocca suture."

(Signed) Brendan D. Leahey,
Lowell, Massachusetts.

Request for information

Editor,
American Journal of Ophthalmology:

Finding the following reference in my reading, I wondered if there was further light in the literature or in medical annals on the condition of the eyes of the great historian, William H. Prescott. One also wonders if there is anyone who could describe the "writing-case for the blind" which he mentions as "not permit(ting) the writer to see own manuscript." The reference will be found in the preface of Prescott's *History of the Conquest of Mexico:*

"For one thing, before I conclude, I may reasonably ask the reader's indulgence. Owing to the state of my eyes, I have been obliged to use a writing-case made for the blind, which does not permit the writer to see his own manuscript. Nor have I ever corrected, or even read, my own original draft. As the chirography, under these disadvantages, has been too often careless and obscure, occasional errors, even with the uttermost care of my secretary, must have necessarily occurred in the transcription, somewhat increased by the barbarous phraseology imported from my Mexican authorities . . ." (1843).

(Signed) Victor C. Rambo,
Philadelphia, Pennsylvania.

(Editor's note: If any reader can answer these questions, will he please communicate with Dr. Victor C. Rambo, 156 West School Lane, Philadelphia 44.)

BOOK REVIEWS

Documenta Ophthalmologica: Advances in Ophthalmology. Edited by von Bahr, Ten Doesschate, Fischer-von Bünau, J. François, Goldmann, Lo Cascio, Müller, Nordmann, Schaeffer, Sorsby. 'S-Gravenhage, W. Junk, 1958. Volume XII, 365 pages, with supplement. Price: $25.50.

This volume of the widely known and useful *Documenta Ophthalmologica,* consists of three monographs, two in English. The first is that of P. C. Donders of Groningen on "Eales's disease." It is a most comprehensive study of 100 cases, with historical background and a discussion of treatment with diathermy coagulation. In so far as etiology is concerned nothing new has been added. The author cannot discount the tuberculosis theory but believes that in time allergy may be found to be an important factor. Thirty-seven of his patients were treated with surface coagulation which was successful in 64.8 percent of the cases. It is not desirable to intervene in advanced cases. Light coagulation offers great possibilities. (Summaries in French, German, extensive bibliography.)

The second paper is by H. O. Depner, Nancy, on the "Present status of tonography" (in French). This is a thoughtful and highly technical study of the dynamics and

formulas of tonography. He concludes that the tonographic outflow resistance is not entirely mechanic or "dead" but contains a "living neurovascular" fraction of about five to 15 percent. Numerous tonographies on the same eye are close to reality. There is a wide margin of error in single tonographies. The method is restricted to special cases (1) low tension and hypersecretion glaucomas and (2) the early prognosis of the capacity of outflow following glaucoma surgery. (Summaries in English, German. Extensive bibliography.)

The third article, "Mechanisms of reflex dilatation of the pupil," is by Irene E. Loewenfeld of New York, working in the laboratory of Otto Lowenstein who writes a short preface. Dr. Loewenfeld's work is most extensive and elaborate, beginning with a long historical study of the background and ending with a detailed analysis of her experimental work. The supplement consists of reproductions of her working sheets of analysis of her data. I find it impossible here even to summarize her summary. (Extensive bibliography, summaries in English, French, German.) Her paper will bring a lot of pleasure to the ocular physiologists.

The volumes of *Documenta Ophthalmologica* are indispensable tools, particularly for the research worker.

Derrick Vail.

A Documental Study of the Fundus Oculi: Volume I. Alterations in the Retinal Circulation. By Antonio Clerici, M.D. Milano, Italy, A. Fattorini, 1957. 133 photographs, 210 pages, 136 references. Price: Not listed.

This textbook can be divided into a series of nine sections, dealing in numerical order with spasmatic conditions of the retinal vessels, retinal hemorrhages, exudates, hypertensive retinopathy, venus occlusion, arterial occlusion, diabetic retinopathy, and renal retinopathy.

The story of these conditions is told quite vividly by means of photographs. The photographs are large and are well reproduced and certainly provide, primarily for the beginner, a graphic representation of retinal conditions. The section covering hypertensive retinopathy and arteriosclerosis was particularly interesting. The author prefers the Scheie classification in which the findings of hypertension are separated from those of arteriosclerosis. The section covering retinal hemorrhages is particularly well illustrated. He gives us a photographic history of a preretinal hemorrhage from its inception through its various stages of absorption and complete resolution two years later.

The book would certainly be of great help to medical students who are attempting to learn funduscopy.

Joseph E. Alfano.

Rare Ocular Tumors. By Konstantin Pascheff, M.D. Sofia. (German translation edited by Ruth Börner, M.D.) Berlin, Akademie Verlag, 1958. 141 pages, 98 illustrations, 7 tables. Price: Not listed.

Rare Ocular Tumors and Ocular Cancer in Bulgaria, originally published in 1952, and *Plasmocytic and Lymphocytic Hyperplasias and Tumors of the Conjunctiva,* originally published in 1951, are combined in a single volume in this translation from Bulgarian into German and edited by Börner.

As a product from the pen of the Nestor among Bulgarian ophthalmologists, this work deserves respect. The cases described were all examined by Pascheff during the many years of his directorship in the eye clinic which he founded in Sofia. Yet one wonders about the title of a book on "rare" tumors which includes examples of malignant melanoma of the choroid and retinoblastoma. Most tumors are represented by one, or, at the most, two cases. The meager statistical data are of little help in an evaluation of the frequency with which some of the rare tumors occur. Also, "tumor" is used in the widest sense of the word, including not only

instances of syphiloma but also parasitic cysts.

In variance to our present tendency of simplifying the nomenclature, Pascheff clings to subdivisons and even attempts to introduce new subheadings. A term like melanoneurophoroblastoma is quite descriptive but is bound to create confusion. A case of cystic basal-cell carcinoma is presented as cylindroma. This term, originally introduced by Billroth, is used by some authors for certain types of carcinoma of the lacrimal gland—an example of how such subdivisions add to the difficulties in our understanding of the variety of tumors.

There is one example of this rarest of all intraocular tumors: one originating from the pigment layer of the retina. Yet, from the description of the case and the microphotograph it would be most difficult to differentiate it from an ordinary epithelioid cell type of malignant melanoma of the choroid.

The second part of the book concerns itself with a re-evaluation of Pascheff's original contributions to the plasmocytic and lymphocytic hyperplasias of the conjunctiva. He has not abandoned his original concept that plasmoma is an inflammatory growth sui generis and is in no causal relationship to trachoma—in spite of the fact that seven of his 10 cases suffered from trachoma.

In summary, this book should be considered a document of personal reminiscences of one of ophthalmology's great names even though it is quite dated in many respects. It adds little to our present knowledge of the subject and in no way challenges the standard texts of the tumors of the eye and its adnexa.

Stefan Van Wien.

War Blinded Veterans in a Postwar Setting. Published by Veterans Administration, 1958. 260 pages, 88 tables. Price: $1.50.

How have the veterans who were blinded in World War II and the Korean Police Action fared since the end of hostilities and the end of their blind training? This volume prepared by the Social Work Service and the Physical Medicine and Rehabilitation Service of the Department of Medicine and Surgery of the Veterans Administration is a noteworthy effort to find out.

It will be recalled that toward the end of World War II our government established training centers for the blinded service men at Avon Old Farms Convalescent Hospital, Valley Forge Army General Hospital, Dibble Army General Hospital, and Philadelphia Naval Hospital. Shortly after the conclusion of the war the Blind Rehabilitation Unit in the Veterans Administration Hospital, Hines, Illinois, was established "to serve the needs of veterans suffering postwar loss of sight from old injuries incurred in service and also service men losing their sight in such future military action as might occur." This unit was an active and going concern when the Korean War broke out and was therefore able to fulfill its obligations to the blinded service men of that episode without delay. The training program is superb and most efficient.

A follow-up study of the veterans who had passed through these centers was initiated in about 1953. Voluminous studies were made by a host of dedicated workers. The fruits of their labors are detailed in this book, which of its nature is statistical and therefore does not lend itself readily to review. It is worthy of careful attention by all those interested, and this includes ophthalmologists, in the reaction and lives of the blind.

At the outset, the reader is impressed with the warmth, compassion, and sympathetic approach of the many workers who have produced this report. All of us should read and ponder Chapter II, "Purpose of survey report." Here after discussing the conflicts between the seeing and the blind, and in the blind man himself, the chapter closes with this paragraph: "It is always well to keep in mind the fact that the inward being of the individual who has been blinded as an adult is often a battleground between the whole

man and the man with very great limitations within one body, meeting in sharp conflict for the first time. It is with the resolution of this conflict that one phase of blind rehabilitation is concerned. A study of this resolution and how more favorable conditions may be created for it is the essential integrating purpose of this survey."

At the end of the study, Chapter XV, "Conclusion," ably sums up the findings of the study. It is too long to quote here, but I should like to reveal the "final note": "Finally, it may be said that, despite many hazards of their day and time, the group of citizens studied have emerged not as a weak, disturbing element in social structure, but as an inspiriting human phenomenon, who serve and enrich their country and their world" . . .

The Veterans Administration and particularly those who were responsible for the excellent and heart-warming report are to be congratulated. The subject is a most difficult one and the statistics must have been arduous to an extreme to gather and compile. The analysis of the data is most masterly and we all should be very proud of our Veterans Administration.

Derrick Vail.

OPHTHALMIC FITTING AND ADJUSTING. By Frank Kozol, B.S., O.D. Philadelphia, Chilton Co., 1958. 155 pages, 214 figures, index. Price: $6.50.

Fitting and adjusting procedures vary according to the ever-changing mechanical structure and styling of frames and mountings. This thoroughly up-to-date manual stresses painstaking accuracy; many times a slight movement of a pad or temple may make the difference between the patient's being able to wear a correction in comfort or not being able to tolerate it at all. Hence the ophthalmologist should check not only the accuracy of the lenses but the fit of the frame. Knowing the needs of the patient, he can then suggest what alterations may be required. A reading of this book will be most helpful in determining what can and should be done.

James E. Lebensohn.

SYSTEMIC OPHTHALMOLOGY. By Arnold Sorsby. London, Butterworth & Co.; St. Louis, C. V. Mosby, 1958. Second edition. 664 pages, 277 illustrations, 24 color plates, selected bibliography, index. Price: $25.00.

Prof. Sorsby, F.R.C.S., has found it necessary to bring out a second edition of his popular book which first appeared in 1951, not only because of the demand for it but also because in these short seven years further important advances in ophthalmology in relation to general medicine have occurred. Thus he has revised his text and has incorporated entirely new chapters. There are 34 contributors, some old, some new. While there are a few less illustrations than in the first edition, the loss is not felt. It has been a valuable book for study and reference and the new edition is an improvement.

Derrick Vail.

ABSTRACT DEPARTMENT

EDITED BY DR. F. HERBERT HAESSLER

Abstracts are classified under the divisions listed below. It must be remembered that any given paper may belong to several divisions of ophthalmology, although here it is mentioned only in one. Not all of the headings will necessarily be found in any one issue of the Journal.

CLASSIFICATION

1. Anatomy, embryology, and comparative ophthalmology
2. General pathology, bacteriology, immunology
3. Vegetative physiology, biochemistry, pharmacology, toxicology
4. Physiologic optics, refraction, color vision
5. Diagnosis and therapy
6. Ocular motility
7. Conjunctiva, cornea, sclera
8. Uvea, sympathetic disease, aqueous
9. Glaucoma and ocular tension
10. Crystalline lens
11. Retina and vitreous
12. Optic nerve and chiasm
13. Neuro-ophthalmology
14. Eyeball, orbit, sinuses
15. Eyelids, lacrimal apparatus
16. Tumors
17. Injuries
18. Systemic disease and parasites
19. Congenital deformities, heredity
20. Hygiene, sociology, education, and history

1

ANATOMY, EMBRYOLOGY, AND COMPARATIVE OPHTHALMOLOGY

Vrabec, F. **Studies on the corneal and trabecular endothelium. I. Cement substance of the corneal endothelium.** Brit. J. Ophth. **42**:529-534, Sept., 1958.

It was found that the intercellular cement substance corresponds to previous classical descriptions in that it forms coarse granules of irregular shape and size and was found to be a somewhat different substance between the cells from that which covers the cells. The material between the cells took up the silver impregnation while that covering the cells did not. This latter substance is believed to differ from the previously described cement and to be secreted by the endothelial cells themselves. (4 figures, 6 references) Morris Kaplan.

3

VEGETATIVE PHYSIOLOGY, BIOCHEMISTRY, PHARMACOLOGY, TOXICOLOGY

Ainslie, D. and Henderson, W. G. **Soframycin. Its penetration into the eye and its effect upon experimentally produced staph. Aureus and ps. pyocyania corneal infections.** Brit. J. Ophth. **42**:513-517, Sept., 1958.

In the treatment of intraocular infection by subconjunctival injection, a new antibiotic, soframycin, seems to give much promise. It is easily soluble in water and is stable in solution. Adequate studies by animal injection have indicated that it is almost completely nonirritating to tissues. In vivo, pseudomonas pyocyanea, staphylococci and 60 strains of coliform organisms were found to be satisfactorily sensitive to it. Experimental corneal infections in animals cleared rapidly after subconjunctical injections. Clinical use has as yet been quite limited. (8 references) Morris Kaplan.

Bonavolonta, A. **The effects of experidine phosphatebenzoicarbinolotrimethyl acetate with ascorbic acid in experimental uveitis.** Boll. d'ocul. **37**:365-377, May, 1958.

The author believes that this compound is at least as useful as cortisone in the treatment of experimental uveitis in rabbits. (10 references) Joseph E. Alfano.

Campbell, D. A., Renner, N. E. A. and Tonks, E. **Effects of Diamox on plasma**

bicarbonate and on the electrolyte balance in relation to intraocular pressure in man. Brit. J. Ophth. **42**:732-738, Dec., 1958.

Observations are recorded after a single dose of 250 mg. of Diamox in a series of 10 glaucoma patients. Sodium and fluid intake was limited in order to avoid blood changes. The results indicate that the immediate drop in intraocular pressure is not related to the small reduction in plasma bicarbonate noted. Under continuous Diamox therapy the chloride concentration in the blood falls during the first few hours and then rises. It is believed that the reduction of sodium and chloride ions is probably more significant than that of the other ions in relation to intraocular pressure. (3 figures, 1 table, 27 references) Lawrence L. Garner.

de Conciliis, U. **Action of butazolidine on the retinal and lens metabolism.** Arch. di ottal. **62**:209-214, May-June, 1958.

3,5-dioxy-1,2-diphenyl-4-n-butyl-pyrazolidine has a steroid-like effect on inflammation. The mode of action is not clear. In rabbits the intravenous injection of 150 mg. per kg. for 35 days caused no change in oxygen consumption or anaerobic glycolysis of the lenses and retinas. (3 figures, 2 tables, 6 references)
Paul W. Miles.

de Conciliis, U. **Catalysis activity of normal and cataractous lenses in humans and rabbits.** Arch. di ottal. **62**:189-195, May-June, 1958.

The human eyes used in these experiments were obtained from surgical enucleations and autopsies. Uveitis was induced in rabbit eyes to produce the unilateral cataracts used in the analyses. Titrimetric and manometric methods showed catalase activity to be less in cataractous lenses than in the normal. (5 tables, 3 references)
Paul W. Miles.

De Simone, Silvio. **Lens glutathione in ditizone experimental diabetes.** Arch. di ottal. **62**:223-231, May-June, 1958.

In three normal rabbits the glutathione content of the lens was 400 mg. percent. Diabetes, which developed in the rabbits 72 hours after the intravenous injection of ditizone, reduced the glutathione to 280 mg. percent. After 15 days it fell to 141 mg. percent, and after 30 days it became only 70 mg. percent. The importance of lens glutathione to diabetic cataract is discussed. (4 tables, 25 references)
Paul W. Miles.

Di Martino, C. **On tests of PCR in experimental ocular pathology.** Arch. di ottal. **62**:215-222, May-June, 1958.

The PCR test determines the proportion of a certain protein in serum globulin by means of paper electrophoresis. The PCR, like the sedimentation rate, has considerable sensitivity to the relative activity of a systemic disease. It has been used in rheumatic fever to determine the success of treatment.

Experiments were made on 24 rabbits comparing the PCR to the sedimentation rate in ocular inflammation of various degrees produced by cauterizing or burning the cornea or by injecting staphylococci into the anterior chamber. The two tests were found to be equally sensitive. (3 tables, 1 reference) Paul W. Miles.

Huggert, A. **Experimentally induced increase in the intraocular pressure in the rabbit eye.** Acta ophth. **36**:750-760, 1958.

India ink was injected into the anterior chamber of 14 rabbit eyes; a brief increase of the intraocular pressure was followed by a slight reduction accompanied by increased resistance to outflow and a reduction in the aqueous formation. Ligation of the vessels at the posterior pole in eight eyes resulted in an immediate marked increase in pressure followed by a period of moderate increase. Appreciable

changes in rigidity made it difficult to estimate the magnitude of the changes. (1 table, 14 references) John J. Stern.

Lewis, J. G. and Stephens, P. J. **Tear glucose in diabetics.** Brit. J. Ophth. **42**: 754-748, Dec., 1958.

A Clinistix was used to touch the tears in the 119 of 200 known diabetics who had blood sugar levels above 160 mg./100 ml. In 85 percent of them the tear glucose level was sufficient to produce a positive reaction. The 15 percent in whom no color reaction occurred indicate that a negative reaction cannot be used as evidence that diabetes is absent. (1 figure, 1 table, 5 references)

Lawrence L. Garner.

Linnér, E. and Törnquist, R. **Facility of outflow in shallow anterior chamber.** Acta ophth. **36**:687-692, 1958.

Twenty-one patients with acute glaucoma and shallow anterior chamber were studied. A second group consisted of normal individuals, mostly siblings of the patients. No correlation was found between the facility of aqueous outflow and chamber depth. (3 figures, 1 table 14 references) John J. Stern.

Ohashi, Kohei. **Clinical observations on aqueous flow and resistance to flow using the bulbar pressure test.** Klin. Monatsbl. f. Augenh. **133**:653-661, 1958.

In 30 normal eyes the author measured the ocular tension with the Schiøtz tonometer and the pressure in the anterior ciliary and vorticose veins with the ophthalmodynamometer. The difference between the ocular tension and the venous pressure is much higher in glaucomatous eyes (20 to 22 mm. Hg) than in normal ones (8 to 13 mm. Hg). The outflow through the anterior channels, mainly Schlemm's canal and the anterior ciliary veins, is irregular and inconstant and can be regarded as a safety valve. The outflow through the posterior channels, mainly the vorticose veins, is even and continuous and caused by differences in osmotic pressure. (1 figure, 6 tables, 26 references) Frederick C. Blodi.

Oksala, A. and Lehtinen, A. **Absorption of ultrasound in the aqueous humor, lens and vitreous body.** Acta ophth. **36**:761-768, 1958.

The absorption of ultrasound in normal aqueous and vitreous is equal to that in distilled water. Absorption in the lens is considerably higher. (4 figures, 1 table, 10 references) John J. Stern.

Oksala, A. and Lehtinen, A. **Measurement of the velocity of sound in some parts of the eye.** Acta ophth. **36**:633-639, 1958.

Using an interferometer with an ultrasonic apparatus, the velocity of sound in some tissues of the eye was measured, with the following results: sclera 1,630 m./sec., cornea 1,550, aqueous 1,495, vitreous 1,495 and lens 1,650. These differences explain the observations of echoes from the border surfaces within the eye. (5 figures, 8 references) John J. Stern.

Rexed, Ursula. **The pH of the lachrymal fluid determined by a capillary microglass electrode.** Acta ophth. **36**:711-718, 1958.

There is a considerable difference in the results of previous attempts to determine the pH of the tears. The author devised a method by which the tears are collected directly at the orifices of the lacrymal ducts and shielded from exposure to air. Lacrimation was provoked by touching the cornea with a piece of filter paper. A microglass electrode and reference buffer solutions were used, and the method was shown to give an experimental error or ± 0.02. The pH in 100 subjects varied between 7.1 and 7.99. The mean is 7.49 ± 0.02. The standard deviation is 0.16. (3 figures, 3 tables, 16 references)

John J. Stern.

Rosin, A., Lorian, V. and Ratiu, E. **Cetavlon in ophthalmology.** Ann. d'ocul. **191**:598-600, Aug., 1958.

Cetavlon is a new disinfectant and bacteriocide. It is cetyltrimethylammonium bromide and has great surface activity. The authors use it in 1/2000 solution and 1/500 ointment. A weaker solution (1/5000) is used for maintenance of prostheses and contact lenses. Almost all cases of conjunctivitis respond rapidly to treatment in 4 to 11 days. (2 references)

David Shoch.

Sorsby, A. and Nakajima, A. **Experimental degeneration of the retina. IV. Diaminodiphenoxyalkanes as inducing agents.** Brit. J. Ophth. **42**:563-571, Sept., 1958.

Clinical use of the new schistosomicidal drugs, the diaminodiphenoxyalkanes, has shown that the ingestion of some of them may be followed by retinal degeneration similar to retinitis pigmentosa. In this study the effects of the drug in the rabbit are investigated. Various combinations of these amines were used in experiments similar to those previously reported. Of the seven drugs injected intravenously, six were followed by marked retinal degeneration and the seventh produced slight and inconclusive change. Within a few days the retinas became edematous with narrowing of the vessels and much pigment deposition. The nature of the action of these carbon groups is not yet understood. (2 figures, 3 tables, 16 references)

Morris Kaplan.

Sorsby, A. and Nakajima, A. **Experimental degeneration of the retina. III. Inhibitors of glycolysis and of respiration as inducing agents.** Brit. J. Ophth. **42**: 558-562, Sept., 1958.

The electrical activity of the mammalian retina is dependent directly on its high glycolytic activity and therefore it might be assumed that if this glycolysis is interfered with degeneration of the retina should ensue. In this study glycolysis and respiration inhibitors are used in experimental animals to determine the validity of this assumption. The techniques of these experiments have been described in previous articles. A number of agents were used which either inhibited glycolysis or respiration but none of these resulted in retinal damage. However, when some of these were injected in combination, marked degenerative changes resulted. This effect was particularly marked when the combination used was sodium cyanide and sodium fluoride. This suggests that physiology of the retina is highly dependent on glycolysis and respiration. (2 figures, 2 tables, 5 references)

Morris Kaplan.

4

PHYSIOLOGIC OPTICS, REFRACTION, COLOR VISION

Cambiaggi, A. and Cerri, A. **Studies on the influences of acoustic stimulation of dark adaptation of the normal eye.** Boll. d'ocul. **37**:431-439, June, 1958.

The effect of sound stimulation on the threshold of dark adaptation was studied in 60 normal eyes in which the pupil was of normal size, and after dilatation of the pupil and constriction of the pupil with and without a stenopasic slit. It was found that the threshold of dark adaptation was improved by the acoustic stimulation only when the pupil had been dilated. (7 tables, 11 references)

Joseph E. Alfano.

Gramberg-Danielsen, B. **The ophthalmologist's view of dysmegalopsia and metamorphopsia.** Ophthalmologica **136**: 12-39, July, 1958.

The author surveys his observations of dysmegalopsia and discusses the numerous theories of origin of this rarely recognized disturbance. He distinguishes dysmegalopsy in a narrow sense, which is really false peripheral perception, from

the dysmegalopsy that is a faulty interpretation of an optical image which is still true in the first neurone. The causes of this disturbance are reviewed and one more is added to the list, namely transient angioneurotic edema. The author points out that one of the dysmegalophanias, hemidysmegalophania, is important for the early recognition of brain tumor. He also discusses the classification of errors of judgment of size difference of central origin. (157 references)

F. H. Haessler.

Masci, E. **Magnification in subnormal vision.** Boll. d'ocul. **37**:255-282, April, 1958.

The author feels that many patients with subnormal vision can be rehabilitated by proper magnifying lenses. He reviews the principles and efficiency of both simple and telescopic magnification devices. The magnification of the retinal image is directly proportional to the near point to the eye at which the object is held. A greater number of retinal receptors is stimulated by a larger image. (6 figures, 27 references)

Joseph E. Alfano.

Raubitschek, E. **The determination of astigmatism with the aid of the Astikorrekt.** Acta ophth. **36**:719-726, 1958.

Most differences of opinion and interpretation regarding cylinder retinoscopy are based on semantics. The Astikorrekt together with a Purvis streak retinoscope is a useful device for retinoscopy and for subjective vision tests. (The Astikorrekt is a device constructed by the author but not described in this article). (1 figure, 13 references) John J. Stern.

Tiberi, G. F. **The median curve and physiological limits of retinal dark adaptation.** Arch. di ottal. **62**:201-207, May-June, 1958.

This paper is a simple description of the technique in the use of the Goldmann-Weekers adaptometer. Statistical formulas are given to produce the median and the limits. (1 figure, 6 references)

Paul W. Miles.

5

DIAGNOSIS AND THERAPY

Ambos, E. **The "devil's hook", a forgotten fixation instrument.** Klin. Monatsbl. f. Augenh. **133**:719-720, 1958.

This very old instrument was devised by Lüer for fixating the eyeball. It consists of a double hook whose slightly screw-shaped and crossed claws pierce into the sclera. (1 figure, 1 reference)

Frederick C. Blodi.

Boeke, Wilhelm. **The problem of an allergic opacification of corneal transplants.** Klin. Monatsbl. f. Augenh. **133**: 645-653, 1958.

A comparatively late opacification of the graft (two to five weeks after the operation) is generally believed to be due to an allergic reaction or to some other immunobiologic process. The author believes that this is a rare occurrence and perhaps altogether questionable. It certainly can not be caused by an organ-specific process, as bilateral grafts often remain transparent. (1 figure, 18 references). Frederick C. Blodi.

Dorello, U. and Dell'Aquila, A. **Postoperative astigmatism following cataract and glaucoma surgery.** Boll. d'ocul. **37**: 241-253, April, 1958.

The authors found a change in the axis of the astigmatism toward the 180 degree meridian after 60 intracapsular cataract extractions and 50 iridencleises for glaucoma. The changes were more noticeable after cataract extraction and there was little of no difference between the two eyes. The average degree of astigmatism was 2.3 diopters after cataracts and 0.9

diopters after iridencleisis. (10 tables, 44 references) Joseph E. Alfano.

Gedda, L. **A test of twins: the identity of reactions to audiovisual stimulation.** Ann. d'ocul. **191**:752-766, Oct., 1958.

The author took two groups of twins, one identical the other fraternal and subjected the two groups to various tests. He found that head and limb reactions to visual stimulation in the cinema were the same in identical twins but not in fraternal twins. The identical twins could more easily recognize each other's voice than fraternal twins. Recognition of numbers was dependent more on the level of education than on heredity. An auxiliary finding of interest was that prolonged watching of TV seemed to produce an esophoria. David Shoch.

Katclyanski, E. O. **Intravenous injection of novocaine in certain eye diseases.** Vestnik oftal. **2**:33-41, March-April, 1958.

Novocaine was injected intravenously according to the following scheme: a 0.5 percent solution in double distilled water daily in increasing doses of 5, 6, 7, 8, 9, and 10 cc., following which a 1-percent solution was injected daily in similar doses, and finally 10 cc. of a 1-percent solution was given daily for five days. This form of therapy was administered to 50 patients with various corneal diseases, 23 patients with iridocyclitis of various origin, and 47 patients with perforations and contusions of the globe and burns of the cornea and conjunctiva. Complications of the treatment were minor and occurred in only a few patients. Pain accompanying the ocular condition was diminished or removed and the clinical course definitely improved in 82 percent of the patients. (1 table, 17 references)

Victor Goodside.

Montaldi, Mario. **Use of subconjunctival and retrobulbar prednisolone in ocular affections.** Arch. di ottal. **62**:175-188. May-June, 1958.

No advantage was found in the retrobulbar use of prednisolone over subconjunctival injection in diseases of the posterior pole. The prednisolone was used in 10 to 15 mg. doses in 0.5 cc. of water, every two or three days. It was well tolerated and was found to be beneficial in deep metaherpetic keratitis (10 cases), bullous keratitis (2 cases), striate keratitis (10 cases), edema after keratoplasty (3 cases), postoperative iridocyclitis (5 cases), traumatic iridocyclitis (16 cases), acute iridocyclitis (16 cases), chronic iridocyclitis (20 cases), corneal ulcer with hypopyon (9 cases), and perforating corneal ulcer (14 cases). (29 references)

Paul W. Miles.

Mylius, K. **General principles for pre- and postoperative care in eye operations.** Klin. Monatsbl. f. Augenh. **133**:639-645, 1958.

Each patient should be checked for thrombophlebitis of the legs. A chronic bronchitis should be treated before the operation. Early ambulation is advised. (1 table) Frederick C. Blodi.

Niedermeier, Siegfried. **The local administration of glucose in ophthalmology.** Klin. Monatsbl. f. Augenh. **133**:710-711, 1958.

A new ointment combining dextrose with a sulfonamide may be of value in treating corneal wounds, ulcers and inflammations. (7 references)

Frederick C. Blodi.

Pirodda, A. **Some ocular observations during the recent Asian flu epidemic.** Boll. d'ocul. **37**:471-477, June, 1958.

The author observed two cases of orbital cellulitis during the recent Asian influenza epidemic, one case of exudative iridocyclitis and choroiditis, and a considerable increase in the number of cases

of acute congestive glaucoma. The possibility of a relationship existing between the influenza and these eye conditions is discussed. (1 figure, 1 table, 19 references)
Joseph E. Alfano.

Snodgrass, Marjory B. **Retinal vascular micrometry by the use of a measuring ophthalmoscope.** Brit. J. Ophth. **42**:535-539, Sept., 1958.

A graticule was made to fit into the Keeler measuring ophthalmoscope so that it could be rotated and be focused onto the retina. The instrument was calibrated by frequent examinations and measurements on a series of healthy eyes. (3 figures, 1 table, 8 references)
Morris Kaplan.

Tóth, Z. **Fundus changes in juvenile hypertension.** Klin. Monatsbl. f. Augenh. **133**:670-673, 1958.

In juvenile patients with cardiovascular hypertension the retinal vessels may be tortuous and the disc hyperemic. Tóth discusses the relation of these changes to the neurovegetative system.
Frederick C. Blodi.

Vedel-Jensen, Niels. **General anesthesia for intrabulbar operations.** Acta ophth. **36**:681-686, 1958.

Sixty-six cataract operations were performed under narcodorm-pethidine anesthesia. Narcodorm is the Na salt of β-bromallyl-isopropyl-N-methyl barbituric acid and pethidine is dolantin. Ten of the patients also received tubocuran, a curare preparation. No advantage was demonstrated in the administration of curare, and its use is discouraged because of the possibility of undesirable side effects. (2 tables, 13 references) John J. Stern.

6

OCULAR MOTILITY

Huysmans, J. **Experiences with the technique of stretching the oculomotor muscles.** Ann. d'ocul. **191**:601-608, Aug., 1958.

Two years ago the author described his technique of forcible stretching of muscles for the correction of tropias. This is done after injection of hyaluronidase into the sheaths of the muscle. He reports five cases in which this procedure was employed. Four of the patients had good fusion before the stretching. All extraocular muscles are apparently amenable to treatment and in these five cases the lateral, inferior, and superior rectus muscles were stretched. All patients had postoperative third grade fusion. (5 figures) David Shoch.

Meisner, Rosemarie. **Fusional divergence movements in acute esotropia.** Klin. Monatsbl. f. Augenh. **133**:712-713, 1958.

In a 27-year-old woman an acute esotropia of more than 30 degrees followed a unilateral iritis. When an object was brought toward the eyes a divergence movement resulted which also relieved the diplopia. This movement is explained as an expression of relative fusional divergence. (3 references)
Frederick C. Blodi.

Sokolić, Petar. **Double folding of the eye muscle by simple suturing in operation for squint.** Acta ophth. **36**:646-650, 1958.

Three U-sutures are used to fold the muscle to be shortened. The procedure is described in detail. (1 figure, 6 references) John J. Stern.

Sternberg, A. and Fehér, M. **Five years experience with divergent squint.** Ophthalmologica **136**:1-12, July, 1958.

The authors analyze their findings in 248 patients who had been studied during the years 1950 to 1955. Periodic divergence or divergence excess begins early in life, binocular vision is often preserved and amblyopia is less common. Among

their patients the squint was alternating in 80 percent and monocular in 20 percent. In periodic divergence the fusion faculty is too weak in relation to the position of rest. Permanent divergence begins at various ages. Alternation was half as frequent as in periodic divergence and monocular squint was three times as frequent. (10 tables, 7 references)

F. H. Haessler.

7

CONJUNCTIVA, CORNEA, SCLERA

Andersen, S. R. **Pemphigus of the conjunctiva with oral and cutaneous manifestations.** Acta ophth. **36**:640-645, 1958.

A case is reported and the literature is reviewed (3 figures, 7 references)

John J. Stern.

Collier, L. H., Duke-Elder, S. and Jones, R. **Experimental trachoma produced by cultured virus.** Brit. J. Ophth. **42**:705-720, Dec., 1958.

A blind human volunteer was inoculated with trachoma virus and observed for about seven months. The changes that occurred in the palpebral conjunctiva, histologically as well as clinically, coincided with the typical clinical findings of trachoma. The results would tend to produce further evidence that this virus is in all probability the etiologic agent for trachoma. (11 figures, 5 tables, 7 references)

Lawrence L. Garner.

Negroni, G. **Experimental researches on the corneo-scleral transplantation of the limbic region.** Boll. d'ocul. **37**:459-470, June, 1958.

The author performed a series of corneo-scleral grafts of the limbic area in rabbits. The grafts measured 9 mm. in diameter and included 3 to 4 mm. of cornea. The author reports no unusual complications and suggests further studies. (5 figures, 30 references)

Joseph E. Alfano.

Pannarale, M. R. and Brugo, B. **The use of N, N'dibenzilethylendiamine dipenicillin G. in relapsing hordeola.** Boll. d'ocul. **37**:282-308, April, 1958.

The authors found that in 40 cases of recurrent hordeola which had been resistant to other forms of therapy, the use of N, N'dibenzilethylendiamine G. dipenicillin was effective in all cases. (3 tables, 23 references)

Joseph E. Alfano.

Payrau, P., Bonel, L. and Guyard, M. **Homo—and hetero—grafts of the cornea using grafts presented by lyophilization.** Ann. d'ocul. **191**:636-669, Sept., 1958.

The authors review the various methods of preserving corneas for future use and conclude that chilling and desiccation are best for preserving the structure of the cornea. They describe their technique for preparing such grafts. The enucleated globe, after preliminary preparation, is plunged into a bath of alcohol and solid carbon dioxide. The globe is then dehydrated at a temperature of -0.50 degrees C. for 18 to 20 hours. The flask is then sealed in an atmosphere of nitrogen and may be kept indefinitely. When needed the eye is rehydrated with a physiologic salt solution. These grafts are totally nonviable but have practically an unaltered physical structure. The authors feel that they have lost their antigenic properties and have unquestioned sterility.

The authors report 119 keratoplasties done for 29 human patients. Experience with penetrating homografts is still limited but lamellar homografts do very well and remain transparent. Lamellar heterografts from dog to man seem to do well but similar grafts from pig to man do very poorly. The authors are not yet ready to advise the use of heterografts but they feel that their technique of lyophilization makes homografts and heterografts more alike. (23 figures, 44 references)

David Shoch.

Sedan, J., Farnarier, G. and Ferrand, G. **Contribution to the study of senile corneal anesthesia.** Ann. d'ocul. **191**:736-751, Oct., 1958.

The authors used Franceschetti's kerato-anesthesiometer to measure the corneal sensitivity of 920 patients over the age of 65. They found that generally up to the age of 70 years there is very little diminution in the sensitivity of the central portion of the cornea. Between 70 and 75 years of age central corneal sensation remains essentially normal but there is a marked diminution of the sensitivity at the limbus and particularly of the superior corneal arc. Over the age of 75 years there is, in addition, a marked diminution in central corneal sensitivity.

Generally, other ocular abnormalities do not influence corneal sensation except in the case of patients with cataracts where there is a diminuation in central corneal sensation. Also patients with latent glaucoma seem to have a diminution of the sensation of the superior corneal arc. Systemic diseases seem to play little part in corneal sensation. The most marked changes occur in patients with corneal disease such as arcus senilis, limbal sclerosis and vascular changes at the limbus. In many cases it was felt that the difference in corneal sensation preceded generalized senescence. (7 tables, 39 references) David Shoch.

Smith, C. H. **Accidental laboratory infection with trachoma.** Brit. J. Ophth. **42**: 721-722, Dec., 1958.

While inoculating chick embryo yolk-sacs with trachoma virus some of the infected emulsion accidentally splashed into the face of the operator. No particular cleansing procedure was applied to the eyes and five days later irritation was noted in one eye which within 24 hours had progressed to an intense conjunctivitis. Subsequent tests revealed the presence of a single inclusion body. Therapy with aureomycin and achromycin was not effective and penicillin was substituted. It is of interest to note that the virus had been freeze-dried and transported from China to England, then passed through 23 eggs and was still able to maintain the degree of virulence noted. (4 references) Lawrence L. Garner.

Uzelac, Vladislav S. **Gastro-intestinal extract in the treatment of corneal epithelial dystrophies.** Acta ophth. **36**:630-632, 1958.

A number of albino rats were placed on a diet intended to produce degenerative changes of the cornea. A gastro-intestinal extract was injected intramuscularly daily. Histological examination showed unmistakable regeneration compared with untreated controls. (2 figures, 2 references) John J. Stern.

Vancea, P. **Rodent ulcer of the cornea treated by diathermy coagulation.** Ann. d'ocul. **191**:767-771, Oct., 1958.

The author reports the case of a 52-year-old man with a three months' history of pain in the left eye. Examination showed a typical rodent ulcer in the inferior portion of the cornea. All medical and surgical therapy failed to heal it and vascularization continued. At this point diathermy coagulation of the invading edge of the ulcer was used and repeated one week later. Within two weeks the eye had healed. David Shoch.

Verdi, G. P. and Filippone, A. **A case of heredofamiliary corneal degeneration of Reis-Bückler's type.** Boll. d'ocul. **37**:410-430, June, 1958.

The authors describe a heredofamilial dystrophy of the cornea, probably recessive in nature, observed in two siblings. The opacities observed with slitlamp were localized at the level of the epithelium, Bowman's membrane, and in the superficial stroma. There was some loss of

visual acuity, but no alteration of the corneal sensation. Histologic examination of the cornea confirmed the location of the opacities. After reviewing the literature, the authors believe that the cases reported can be included in the group of superficial corneal dystrophies of the Reis-Bückler type in the classification by Franceschetti. (8 figures, 1 table, 49 references) Joseph E. Alfano.

8

UVEA, SYMPATHETIC DISEASE, AQUEOUS

Krahnstoever, Max. **Serous iritis as the first symptom of trachoma.** Klin. Monatsbl. f. Augenh. **133**:720-721, 1958.

The author was accidentally infected with trachoma 38 years ago. A serous iritis was the first symptom.

Frederick C. Blodi.

Oksala, Arvo. **Observations on choroidal detachment by means of ultrasound.** Acta ophth. **36**:651-657, 1958.

Studies of postoperative choroidal detachment with ultrasound equipment indicate that the suprachoroidal space is filled with homogeneous fluid. The method makes it possible to recognize the presence of tumor tissue in the suprachoroidal space. (4 figures, 25 references)

John J. Stern.

Sivasubramaniam, P. **Fundus changes in uveo-meningitis.** Brit. J. Ophth. **42**:759-761, Dec., 1958.

A case of Harada's uveo-meningitis is described with the classical findings including the uveitis and serous retinal detachment associated with a subsequent graying and falling out of hair and a temporary loss of hearing. An additional finding is described, namely angioid streaks which were noted in both fundi after the retinas become reattached. (2 figures, 2 references)

Lawrence P. Garner.

9

GLAUCOMA AND OCULAR TENSION

Andersen, O. Corydon. **Frequency of glaucoma.** Acta ophth. **36**:672-680, 1958.

Among 800 consecutive patients, 13 were found to have definite glaucoma when they were examined with Goldmann's aplanation tonometer. Eight of them had no complaints and no other clinical signs. (1 figure, 2 tables, 13 references) John J. Stern.

Batarchukov, R. A. **Trephanocyclodialysis in glaucoma.** Vestnik oftal. **1**:10-14, Jan.-Feb., 1958.

The operation of trephanocyclodialysis was carried out on 85 patients (97 eyes), of whom 64 had a congestive form of glucoma. A conjunctional flap was turned down 8 to 9 mm. from the limbus in the upper outer quardrant and a 1.5 mm. trephine opening in the sclera was made 5 to 6 mm. from the limbus. A cyclodialysis spatula was inserted into the opening and pushed into the anterior chamber to produce dialysis of the ciliary body in a wide arc. Postoperatively a full normalization of pathologic intraocular pressure was noted in 86 percent of the patients, a considerable lowering in 13 percent and an increase in intraocular pressure was found in two patients. In 40 patients observation extended from one year to three years and nine months; normal tension was found in 36 patients and in four tension remained elevated. Visual acuity improved or was unchanged in 21 patients, became somewhat diminished in 11, was greatly diminished in six, and ended in total blindness in two. In 47 out of 97 eyes hyphema occurred postoperatively. Usually this was absorbed in a few days. In two eyes blood had to be removed by anterior chamber puncture. Hyphema was not a determining factor in the outcome of the operation. In eight patients there was postoperative iridocyclitis. (1 table, 1 reference) Victor Goodside.

Kalt, M. and Loisillier, F. **Late infections following fistulizing anti-glaucoma surgery.** Ann. d'ocul. **191**:713-735, Oct., 1958.

The author reports 13 late infections in a series of 464 antiglaucoma operations. This is an incidence of 2.8 percent. This compares well with the figures in the literature. The etiology is almost always an exogenous infection from the lids and other contiguous structures. Of these 13 cases six were "benign", that is, only the conjunctiva and the bleb were involved in the infection. In the remaining seven there was intraocular inflammation with hypopyon. The benign cases all cleared promptly with antibiotic therapy. Of the seven serious cases five were cured with intensive local and systemic therapy although in four of them the increased intraocular pressure was permanent. In the remaining two cases the patient became blind. David Shoch.

Leydhecker, W., Akiyama K. and Neumann, H. G. **The intraocular pressure of normal human eyes.** Klin. Monatsbl. f. Augenh. **133**:662-670, 1958.

This report is based on the examination of nearly 20,000 eyes. The tonometers were standardized and the 1955 conversion table was used. There was no difference between the right and left eye, nor between the eyes of men and women. There is a slight increase of the average value with age (from 15.5 in the second to 15.7 in the seventh decade.) The normal range was between 7.5 and 24 mm. Hg. That means that a scale reading of 2.5 with the 5.5 gm. weight is definitely abnormal. (3 figures, 3 tables, 16 references) Frederick C. Blodi.

Santoni, A. **Some considerations on the diagnosis and frequency of glaucoma.** Boll. d'ocul. **37**:401-409, June, 1958.

The author calls attention to the difference in the incidence of glaucoma in Sardinia and Umbria. In 50 percent of the cases, particularly those in advanced stages, the diagnosis can be made quite easily from the tonometric readings and evaluation of the fundus. However, in early cases in which there were no fundus changes or field defects, the tonometer readings were often low and misleading. In these cases he feels that the diagnosis can be made by checking the pressure during the evening hours and also by other tests including tonography. (1 figure, 16 references) Joseph E. Alfano.

Törnquist, Ragnar. **Dark-room test on eyes with a shallow anterior chamber.** Acta ophth. **36**:664-671, 1958.

The dark-room test was performed on 37 eyes with angle closure glaucoma which had not been operated upon, 21 healthy eyes of patients with angle closure glaucoma in the other eye, and 100 eyes with a shallow anterior chamber in nonglaucomatous persons. Positive results were obtained in the three groups in 38, 14, and 3 percent, respectively. In the first two groups, more eyes with a good dilatation of the pupil showed an increase in pressure than eyes with a less marked dilatation; however, many eyes, particularly in the normal group, showed no significant rise in pressure with good mydriasis. No correlation between the depth of the anterior chamber and the dark-room test could be shown in any group. (1 figure, 17 references, 4 tables)

John J. Stern.

10

CRYSTALLINE LENS

Barraquer, Joaquin. **Lens extraction with enzymatic zonulolysis.** Klin. Monatsbl. f. Augenh. **133**:609-615, 1958.

Alpha chymotrypsin was obtained from cattle pancreas. It can be preserved in powder form and should be dissolved in water immediately before the operation.

The tolerance to this enzyme was first tested on animals' eyes and on enucleated human eyes. The enzyme never produced any inflammatory reaction, nor did it affect the lens capsule, hyaloid membrane, iris or cornea. Zonulolysis occurred only in human eyes.

The enzyme was then used for cataract extractions and 276 such operations can be reported. The operations are done under general anesthesia. Chymotrypsin is injected (1:5000 solution) behind the iris into the posterior chamber. The zonule is dissolved in two minutes. Iridectomy is done later. Extraction should be done by suction. The anterior chamber is irrigated with acetylcholine to produce miosis. (1 figure) Frederick C. Blodi.

Charamis, J. **New orientation for cataract extraction with the use of chemical substances.** Ann. d'ocul. **191**:627-635, Sept., 1958.

The author reviews Barraquer's work on alpha-chymotrypsin and reports 56 cases in which the enzyme was employed. Both forceps and erisophake were used. There were three ruptured capsules and two eyes in which vitreous was lost. The author states that the vitreous tends to herniate into the anterior chamber more easily and therefore powerful miosis must be effected after the extraction. It is also essential to inject air after closure of the wound. The enzyme has no affect on posterior synechia. (1 table, 15 references) David Shoch.

Custodis, E. **A contribution to the problem of intraocular infections after cataract extraction.** Klin Monatsbl. f. Augenh. **133**: 632-639, 1958.

Within three consecutive days 12 intraocular infections occurred after cataract extraction. With treatment the final visual results were excellent. The infection could be traced to a pilocarpine solution which was injected into the anterior chamber after the extraction. The organism was Escheridia coli. (2 figures) Frederick C. Blodi.

De Rosa, L. **A particular suture method for cataract surgery.** Arch. di ottal. **62**: 197-199, May-June, 1958.

A preplaced corneoscleral-conjunctival suture is described. (2 figures, 4 references) Paul W. Miles.

Moore, J. Gibson. **Cataract extraction under general anaesthesia.** Brit. J. Ophth. **42**:723-725, Dec., 1958.

The use of general anesthesia with an endotracheal tube is described in a series of 172 cataract extractions. The anesthetic used is nitrous oxide and oxygen and intermittent small doses of pethidine are injected as needed by a skilled anesthetist. Corneoscleral sutures must be used because of the slightly increased risk of coughing or vomiting. Complications seemed no greater than under the usual local infiltration techniques. Two cases of coronary thrombosis were noted after the fifth postoperative day, but this is difficult to attribute to the anesthesia. A thorough physical examination should be performed on all elderly candidates for general anesthesia and if no contraindications are present, it affords a safe and comfortable method for both patient and surgeon. (1 table. (6 references) Lawrence L. Garner.

Nordlöw, W. **The course, complications and final results of 324 consecutive cataract extractions.** Acta ophth. **36**:693-710, 1958.

The article itself is a summary and should be consulted in the original. (19 tables, 9 references) John J. Stern.

Remky, H. **Experiences with the enzymatic zonulolysis of Barraquer.** Klin. Monatsbl. f. Augenh. **133**:616-619, 1958.

Remky reports 68 cataract operations

done with enzymatic zonulolysis. The youngest patient was four years old. The younger the patient the longer must the enzyme remain in the posterior chamber. The extraction was done with the forceps, by expression (Smith-Judian method) or by suction. The latter is the method of choice. The greatest advantage of zonulolysis lies in the ease with which a lens can be extracted from the eye of a child or adolescent. The only danger lies in performing the extraction too soon after the application of the enzyme.

Frederick C. Blodi.

Schenk, H. and Papapanos, G. **The problem of dilating the pupil before a cataract extraction.** Klin. Monatsbl. f. Augenh. **133**:625-632, 1958.

Ten elderly patients were tested. In all of them a 1-percent solution of homatropine alone did not satisfactorily dilate the pupil. It was found that a combination of 1-percent homatropine with the sympathomimetic drug Veritol was most advantageous. The dilatation was maximal and the postoperative constriction with eserine was satisfactory. (1 figure, 1 table, 12 references) Frederick C. Blodi.

Seedoriff, H. H. **Two cases of galactose cataract and a biochemical—ophthalmological survey.** Acta ophth. **36**:658-663, 1958.

The literature is reviewed and two cases in brothers are described. (23 references) John J. Stern.

Walser, E. **First experiences with enzymatic zonulolysis in cataract extractions.** Klin. Monastbl. f. Augenh. **133**:619-624, 1958.

Walser reports 57 operations by means of enzymatic zonulolysis, which presents a great step forward in the field of cataract surgery. The enzyme should remain in the posterior chamber for at least five minutes. Frederick C. Blodi.

11

RETINA AND VITREOUS

Balacco, F. and DeTullio, P. **A rare case of pseudo-aneurysm of the retinal vessels.** Boll. d'ocul. **37**:388-394, May, 1958.

The authors present the case of an arteriosclerotic pseudo-aneurysm of a nasal branch of the retinal artery. The aneurysm was visible ophthalmoscopically and there was an isolated central scotoma. After the instillation of a mydriatic (homatropine) in association with a vasoconstrictor (Fenilfar 10 percent), the ophthalmic picture returned to normal and the central scotoma disappeared. (2 figures, 5 references)

Joseph E. Alfano.

Bech, K. and Jensen, O. A. **Racemose hemangioma of the retina.** Acta ophth. **36**:769-781, 1958.

Two cases of racemose hemangioma (arteriovenous aneurysm) of the retina are presented and the literature is reviewed. In one case defects in the visual field developed after arteriography. On the basis of the evidence in the literature the frequent coexistence of retinal and mesencephalic arteriovenous aneurysms, postulated by Wyburn-Mason, is questioned. (2 figures, 1 table, 20 references)

John J. Stern.

Brücker, R. **The treatment of diabetic retinopathy.** Ophthalmologica **136**:46-53, July, 1958.

A certain optimism in the prognosis is justified, even when repeated retinal hemorrhage has occurred, in a patient who is under the care of an experienced internist. The essentials are a diet rich in protein, poor fat and relatively high in carbohydrate, combined with the use of insulin or sulfonylurea or both. Meticulous education of the patient is of great importance. (7 references) F. H. Haessler.

De Simone, S. and de Conciliis, U. **Angioid streaks of the retina, clinical and pathogenic considerations.** Arch. di ottal. **62**:161-174, May-June, 1958.

Angioid streaks of the retina represent only part of a disease involving elastic tissue throughout the body. Pathologically, there are ruptures in Bruch's membrane, with proliferation of pigment, splitting of the choroid, and hemorrhages. Clinical signs include hemeralopia, decrease in central vision, edema of the macula and choroidal sclerosis. In one case reported, a man, aged 51 years, had a blood pressure of 170/100 and a retinal artery pressure of 80/50. He was acutely ill with early cardiorenal insufficiency. The second case occurred in a man aged 45 years, who had typical pseudoxanthoma elasticum of the skin, and myocardial sclerosis. (2 figures, 43 references)

Paul W. Miles.

Maag, Berthold. **A worm-like parasite in the vitreous of a human eye.** Klin. Monatsbl. f. Augenh. **133**:713-718, 1958.

A motile worm-like structure, probably a filaria, was observed in the vitreous of a 64-year-old woman. Visual acuity was normal. (1 figure, 16 references)

Frederick C. Blodi.

Madroszkiewicz, M. **New method of operation for detachment of the retina with tearing of the macula lutea.** Brit. J. Ophth. **42**:739-748, Dec., 1958.

The author designed a "calibrated electrode" by means of which one can produce electrocoagulation at an accurately measured distance from the corneal limbus without the use of other devices. It is introduced through a small opening which is made in the tendon of the lateral rectus muscle. A diathermy coagulation can be placed near the macula without injury to this structure. Re-adherence of the retina can be achieved without destruction of any remaining cones. (7 figures, 24 references) Lawrence L. Garner.

Orma, H. and Vannas, S. **Observations on the use of PH 203 and Hydergine in some circulatory disturbance of the fundus and vascular retinopathy.** Acta ophth. **36**:734-749, 1958.

PH 203 is a combination of panthesine, a local anesthetic with spasmolytic, antiallergic and ganglion-blocking properties, and Hydergine which inhibits the pressosensitive circulation reflexes. It has a peripheral adreno-sympathicolytic action and a bradycardic and mild sedative effect. Eighteen patients with retinal venous or arterial occlusions and 15 with sclerotic retinopathies were given intravenous infusions of PH 203 for six days, supplemented by intramuscular injections. These injections were continued for up to two weeks; then Hydergine was continued by mouth, together with injections twice weekly, for a period of up to seven months. Fifteen other patients were treated with placebos. No definite conclusions could be arrived at, but unexpectedly good results in some severe cases encourage further investigation. (5 figures, 4 tables, 30 references)

John J. Stern.

Palich-Szántó, Olga. **Calciform retinal detachment.** Ophthalmologica **136**:39-46, July, 1958.

A case is described in which there was a retinal cyst and a large choroidal lesion which exhibited certain characteristics of an inflammatory focus and of coloboma. The changes are ascribed to a disturbance of development of the choroidal fissure. (1 figure, 12 references) F. H. Haessler.

Phillips, Calbert I. **Retinal detachment at the posterior pole.** Brit. J. Ophth. **42**: 749-753, Dec., 1958.

Seven cases of posterior retinal detachment are described which in most instances were limited to the region of a posterior staphyloma as seen in high myopes. In some of these no holes could

be demonstrated. One of the patients was an emmetrope in whom a central detachment with a macular hole developed after disciform degeneration of the macula. It is assumed that the posterior degeneration commonly seen in the highly myopic eye could be an important etiological agent in the formation of central detachments. (3 figures, 7 references)

Lawrence L. Garner.

Pischel, D. K., Clark, G., Kronfeld, P. C., Schepens, C. L. and McDonald, P. R. **Symposium: Scleral resection procedures.** Tr. Am. Acad. Ophth. **62**:189-225, March-April, 1958.

Pischel, Dohrmann K. **Introduction.** pp. 189-193.

Three important conclusions from the previous symposium held six years ago are reiterated: 1. a break in retinal continuity is the fundamental factor in the development of retinal detachments, 2. vitreoretinal adhesions can cause these breaks, and 3. vitreous strands can keep the retina from its bed. In this symposium, the importance of the scleral resection procedures is discussed. The three prime indications are: shrinkage of the retina so it no longer fits the scleral shell; shrinkage of the vitreous; formation of strands in the vitreous.

The panel lists five indication for simple diathermy and five for scleral shortening. Diathermy is indicated when we have: 1. a hole with little if any elevation of the surrounding retina; 2. a detachment involving less than a quadrant, with the holes close together and no evidence of traction; 3. the hole or holes are limited to one quadrant and the retina settles with bed rest; 4. disinsertion less than two positions on the clock and showing preoperative settling; and 5. macular hole with little retinal detachment. In these cases 75 percent successful results should be expected. Scleral resection is advocated when one notes: 1. multiple breaks or one very large tear; 2. a shrunken or inflexible retina; 3. that the retina does not settle with bed rest; 4. degenerative myopia; 5. disinsertions of more than two segments (60°) on the clock. Also included are cases of eyes that show definite vitreous traction strands on the retina, those that occur after known vitreous loss; those requiring secondary operation; and cases of eyes with media which make ordinary observation of the retina impossible. (3 references.)

Clark, Graham. **Mechanical factors in scleral surgery for retinal detachment.** pp. 194-197.

Continued separation may be due to persistent vitreous traction, or to loss of free retinal area through folds, contracture or shrinkage. Since the retina is inextensible the sclera and choroid must be brought to the retina by scleral resection or scleral infolding. The important difference between these latter two groups of procedures is that the resection reduces the scleral area as well as the volume of the vitreous chamber. In the infolding, the scleral area is little changed, but the volume of the vitreous chamber is markedly reduced.

The differences in physical characteristics of the vitreous, retina, choroid and sclera are enumerated.

The scleral resection is advisable where there is loss of retinal area; the infolding is indicated where there is vitreous retraction; and a girdling procedure is indicated when over ½ the globe is involved.

It is mathematically shown that 4 mm. is the widest resection that is ever indicated and that 1.0 to 2 mm. wide resection is enough for most eyes. Similarly, shortening a 75.5 mm. girdling tube by 14.5 mm. would serve for severe shrinkage of the retina.

Kronfeld, Peter C. **The scleral resection operation: current techniques.** pp. 198-205.

The size of the scleral infold or "roll" varies, 1. inversely with the thickness of the sclera removed, 2. inversely with the

intraocular pressure at the time the sutures are tied, 3. inversely with the textural firmness of the sclera, 4. inversely with time, and 5. directly with the depth of the suture bites.

This author feels that the roll should be placed well in front of, or behind the retinal breaks, since he believes that the crest of the roll is not well suited for continued contact between a retinal break and diathermized choroid.

The walling off of a diseased area by double rows of semi-penetrating coagulation to the ora is stressed, and he fully recognizes the supreme importance of draining the subretinal fluid. However, the drainage of fluid should be related to the scleral procedure, so that a normal pressure is maintained throughout.

A typical scleral resection operation is then described in some detail and the discussion is closed with a list of different techniques in current vogue. (14 references)

Schepens, Charles L. **Scleral buckling procedures.** pp. 206-218.

A slightly different point of view from the rest of the panel is presented here. Vitreo-retinal traction, manifested by fixed retinal folds, retinal tears with a rolled convex edge, multiple or giant retinal breaks, and vitreo-retinal bands, is the single evil force to be combatted. This is felt to best be done by the scleral buckle, which must be placed on or posterior to the retinal breaks. This buckle is made higher and more permanent by reinforcement with plastic implantation.

The technique of scleral buckling is described in clear detail, with good description of the various special instruments used. The four standard variations are: 1. buckling with circling polyethylene tube (the commonest), 2. with incomplete tube, 3. without scleral excision, and 4. with a trap door scleral flap.

The circling tube is used where enough fluid can be released to close the buckle and tie the tube. By this method, a buckle 6 mm. high can safely be achieved. The breaks should lie on the anterior slope of the buckle and by good release of fluid the retina should be in contact with the buckle everywhere.

The partial tube is used when there is insufficient fluid for an encircling tube. Scleral buckling without excision is used when the break is so far posterior as to be hardly accessible, and when excessive fluid is lost. The trap door is used in breaks with little or no detachment.

All the techniques are lucidly described and accompanied by excellent drawings. The markedly different pre- and postoperative regimes also are described. (5 figures, 11 references)

McDonald, P. Robb. **Complications of the scleral resection operation.** pp. 219-225.

Complication of lamellar resection are damage to vortex veins, loss of vitreous, and excess slack of retina from too much shortening. When it is a secondary procedure the necrotic sclera may rip, the choroid may be exposed, and vitreous may be lost. The globe may rupture in such areas as the tension is built up by tying sutures on applying diathermy. Similarly, the retinal artery may be occluded by excessive pressure, causing amaurosis. Excessive diathermy can cause massive vitreous retraction, burn holes in the retina, and cause scleral necrosis or choroidal hemorrhage. The use of polyethylene plates can minimize the complications of sutures tearing through tissue, and control of pressure can prevent loss of vitreous and ease difficulties in wound closure.

Postoperative complications include muscle imbalances, uveitis from diathermy, conjunctivitis from foreign material, infections, choroidal detachments which are harmless, glucoma, enophthalmos, and systemic complications such as mental disturbances from patching and medication, and thrombophlebitis.

In scleral buckling one must take care

to seal the retinal breaks, avoid damaging the vortex veins, avoid raising the intraocular pressure too high, and avoid allowing the tubing to penetrate, migrate or irritate. (4 figures) Harry Horwich.

Smith, S. M. and Sorsby, A. **Retinoblastoma: some genetic aspects.** Ann. Human Genetics **23**:50-58, Nov., 1958.

Of 1,217 infants admitted to the Sunshine Home for Blind Babies 59 were blind from bilateral retinoblastoma. An analysis of the recorded data as well as of similar data in the literature shows that most cases are unilateral and sporadic. Only an occasional offspring of such patients is similarly affected; the risk is approximately four percent. When retinoblastoma is unilateral in the parents, a bilateral lesion in the offspring is not uncommon. (6 tables, 31 references)

Irwin E. Gaynon.

Snodgrass, Marjory B. **Ophthalmoscopic examination including measurement of selected retinal arterioles in 15 cases of pre-eclamptic toxaemia.** Brit. J. Ophth. **42**:540-548, Sept., 1958.

In 15 patients it was possible to make pre-eclamptic and postnatal examinations and the findings in each case are described in detail. The vessels chosen for measurement were branches of the nasal and temporal arterioles about three to four disc diameters from the disc. The findings generally followed those previously reported in that there is frequently a narrowing of arterioles in this disease; in about one third of the cases this change was of the extent of a whole grid change. Regular measurement of the caliber of retinal arterioles may be of help in the early diagnosis of pre-eclamptic toxemia. (1 table, 5 references)

Morris Kaplan.

Williams, Cyril E. **Retrolental fibroplasia in association with mental defect.** Brit. J. Ophth. **42**:549-557, Sept., 1958.

In this study of 24 children, 14 of whom were in a home for blind imbeciles, 23 of them were premature and 18 were known to have had oxygen. An analysis of the various lengths of stay in high concentrations of oxygen led to the conclusion that this had no appreciable effect on brain damage. It has been disputed that there is a special association between retrolental fibroplasia and mental deficiency and argued that the brain damage is wholly attributable to the hazards of prematurity. A search of the literature revealed insufficient reports to support this view but it is definitely established that there is a greater incidence of brain defects in the patients with retrolental fibroplasia than in other prematures. (1 figure, 3 tables, 26 references)

Morris Kaplan.

13

NEURO-OPHTHALMOLOGY

Paufique, L., Rougier, J., Wertheimer, J. and Allegre, G. E. **Diagnostic and prognostic value of occular signs in spontaneous meningeal hemorrhages.** Ann. d'ocul. **191**:561-578, Aug., 1958.

The authors list the ocular signs of spontaneous meningeal hemorrhage. These are extra- and intraocular palsies, papilledema, retinal hemorrhages and changes in the visual field. Extraocular palsies occur in from 20 to 60 percent of cases with aneurysms of the circle of Willis. Papilledema and retinal hemorrhages are less common; they occur in about 10 percent of cases but are of serious prognostic import and indicate the necessity for intervention.

David Shoch.

14

EYEBALL, ORBIT, SINUSES

Girard, L. J., Fountain, E. M., Moore, C. M. and Thomas, J. R. **Teratoma of the**

orbit. Tr. Am. Acad. Ophth. **62**:226-233, March-April, 1958.

This is the fifteenth case of a tumor representing another fetus and containing tissues of all three germinal layers. Proptosis, chemosis and enlargement of the orbit were noted four hours after birth. Tarsorrhaphy was performed and the initial diagnosis of orbital encephalocele was made. At age three months an exploratory craniotomy was performed, and a cystic mass containing a tumor the size of an adult thumb was found attached to the optic nerve at the apex of the orbit. At age eighteen months of age the findings were pallor of the disc, attenuation of the retinal vessels and enlargement of the orbit.

Microscopic examination of the tumor revealed a cyst wall lined by squamous epithelium, cartilage, respiratory and intestinal mucous glands, neuronal and glial cells, meninges and a portion of peripheral nerve. (9 figures, 3 references)

Harry Horwich.

Hartmann, Karl. **Orbital fistula in empyema of the frontal sinus.** Klin. Monatsbl. f. Augenh. **133**:706-709, 1958.

Two cases are reported, one supposedly tuberculous, in which an empyema of a frontal sinus broke into the orbit and produced an open fistula. Radical excision of the frontal sinus is advised. (27 references)

Frederick C. Blodi.

Papst, W., Mertens, H. G. and Esslen, E. **Chronic ocular myositis. I. Exophthalmic myositis.** Klin. Monatsbl. f. Augenh. **133**:673-694, 1958.

This first paper on the subject deals with that form of orbital cellulitis which goes together with exophthalmus, palpebral edema, conjunctival injection, chemosis and decreased ocular motility. This form belongs to the large group of pseudotumors of the orbit and must be differentiated from another type of orbital myositis which is not accompanied by exophthalmos and which will be dealt with in another communication.

Seven patients with this disease are described. Three of them became blind on the affected side and optic atrophy resulted. In all of the patients cortical steroids were used with advantage. Electromyograms were of great help in establishing a diagnosis. The tracings were of the myopathic pattern and by this means the condition could be differentiated from neurogenic lesions. In two of the cases muscle tissue could be examined histologically. (9 figures, 24 references)

Frederick C. Blodi.

Tikhomirov, P. R. **Mycosis of the orbit, simulating a malignant neoplasm.** Vestnik oftal. **1**:39-44, Jan.-Feb., 1958.

A 36-year-old woman noted a painless swelling of the right side of her face which later became associated with pain. A doughy swelling appeared at the upper outer angle of the orbit. X-ray study revealed irregular thickening of the roof of the orbit in its medial two-thirds. The mobility of the eye was limited mainly upward and outward. Neurosurgical consultation was obtained and a diagnosis of tumor of the orbit was made and surgical removal was advised. Another X-ray examination disclosed an area of destruction of the orbital roof. A neurologic examination now showed right hyposmia, central paralysis of the right side of the face, inequality of deep reflexes, and later increasing weakness and apathy. A malignant tumor spreading from the retrobulbar area was now considered the most likely diagnosis.

At this time two small swellings of solid consistency appeared in the outer half of the right upper lid. These opened spontaneously, discharging a sanguinopurulent material. Laboratory examination produced no unequivocal diagnosis.

Chemosis of the conjunctiva appeared and exophthalmos increased. Culture from the pus showed nonhemolytic streptococcus. Neurologic manifestations increased and the patient died.

Autopsy showed a defect in the orbital roof through which a mass of yellow-green material extended into the brain. Examination of this material now disclosed the presence of Aspergillus.

Victor Goodside.

Vancéa, P., Triandaf, E., Dobrescu, G. and Cernea, P. **A polycystic teratoma of the orbit.** Ann. d'ocul. **191**:670-679, Sept., 1958.

The authors report the case of a three-day-old child with a corneal ulcer and exophthalmos. The eye was eviscerated and antibiotic treatment started. After the inflammation had subsided a mass was palpable in the orbit and an evisceration of the orbit was done. The mass was found to be attached to the apex of the orbit by a pedicle. The mass itself consisted of numerous cystic cavities.

Microscopic examination showed that it consisted of elements of the central nervous system, ocular tissue, skin and its derivatives, some cartilages, muscle, and blood vessels. (6 figures, 21 references)

David Shoch.

Volpi, U. and Bertoncini, G. **A case of eosinophilic granuloma of the orbit.** Boll. d'ocul. **37**:440-458, June, 1958.

The authors present a case of eosinophilic granuloma of the orbit which manifested itself as a painless swelling in the superior lateral portion of the right orbit and was associated with osteoblastic changes in the right frontal bone. The diagnosis was confirmed by histologic examination. The relationship between this condition and Hand-Christian-Schüller and Letterer-Siwe disease is discussed. (8 figures, 72 references)

Joseph E. Alfano.

15

EYELIDS, LACRIMAL APPARATUS

Ormrod, J. N. **Diverticulum of the lacrimal sac.** Brit. J. Ophth. **42**:526-528, Sept., 1958.

Clinical reports of diverticulum of the lacrimal sac are rare; they may be either congenital, traumatic or inflammatory. A 33-year-old woman with epiphora is described who was found to have a sac diverticulum which pressed on the lacrimal passageway and obstructed it. It was easily removed, with complete cure. (2 figures, 5 references) Morris Kaplan.

Sharma, K. D., Shrivastav, J. B. and Agarwal, S. **Ocular rhinosporidiosis simulating a tumor.** Brit. J. Ophth. 42:572-574, Sept., 1958.

Originally rhinosporidiosis was believed to occur only in the nose but lesions in the conjunctiva and in the lacrimal sac have now been reported. The cause is a spore-bearing fungus which attacks only the male and clinically resembles lymphangioma. Diagnosis is made by histologic study. No treatment is described. A case occurring in an Indian male involving the lower eyelid is presented. (3 figures, 9 references)

Morris Kaplin.

Vänttinen, Sinikka. **Reliability of permeability test and canalicular test in examination of the lacrimal passages.** Acta ophth. **36**:727-733, 1958.

The lacrimal passages in 55 patients with epiphora were examined by inserting a cotton applicator under the inferior turbinate and instilling 5-percent argyrol three times at two minute intervals. At the same time West's canaliculus test was made. In a control series of 34 normal tear passages all but one of the two tests were positive. In 14 cases of dacryocystitis the canaliculus test was positive. In 12 cases of stenosis without suppuration 10 were

positive; this indicates that stenosis without dacryocystitis may be associated with canaliculus insufficiency. In 29 instances of epiphora without clinically apparent cause the permeability was doubtful in four and negative in five. It seems that the lacrimal passages may function inadequately without anatomic cause. (1 table, 10 references) John J. Stern.

Whitwell, John **Denervation of the lacrimal gland.** Brit. J. Ophth. **42**:518-525, Sept., 1958.

Since patients with epiphora are not bothered constantly but rather during times of stress or activity, the logical treatment would be to abolish the reflex lacrimal secretion. Dissections of the cadaver proved that the fibers from the facial nerve are the ones to be divided if this reflex tearing is to be abolished. The nerve, which usually enters at the posterio-inferior corner of the gland, is severed after cutting the lateral canthal ligament and exposing the lacrimal gland in its fossa. Healing is uneventful and much relief of the epiphora can be expected. The operation has been done on six patients with satisfactory results. (4 figures, 1 table, 29 references) Morris Kaplan.

17

INJURIES

Remler, O. **Occupational argyrosis in the photochemical industry.** Klin. Monatsbl. f. Augenh. **133**:695-705, 1958.

The argyrosis was caused by the exposure to silver bromide which constitutes the light-sensitive emulsion of photographic material. Of the 31 patients 14 were also exposed to silver nitrate and 20 patients also had a corneal argyrosis in addition to or independent from a conjunctival pigmentation. The corneal lesion never decreased the visual acuity. The conjunctival argyrosis begins in the semilunar fold and in the caruncle. The interpalpebral fissure is primarily involved. The deposition frequently follows the lymph vessels. (2 figures, 1 table, 7 references) Frederick C. Blodi.

Scharnke, W. **Early Passow operation in burns of the eye.** Muenchener med. Wochnschr. **100**:1651-1653, Oct. 24, 1958.

The author evalutes his experience in the early application of Passow's operation in 58 patients with chemical burns of the eyes. The conjunctiva is incised and undermined to make possible the removal of the toxic transudate. (4 tables, 13 references) F. H. Haessler.

18

SYSTEMIC DISEASE AND PARASITES

Cagianut, B. **The ocular syndrome of macroglobulinemia.** Ann. d'ocul. **191**:579-591, Aug., 1958.

Macroglobulinemia was first described by Waldenstrom in 1944. The complete syndrome consists of a normochromic anemia, hemorrhages into mucous membranes and retina, and increased serum viscosity due to increased macroglobulins. The etiology is unknown but there is a proliferation of lymphoid cells and histioplasmocytes of reticular origin. The diagnosis is made by a sternal marrow smear and by electrophoresis of serum proteins. The symptoms are many and varied. Generally there is an enlarged liver and spleen and a perivascular infiltration of the central nervous system resulting in diverse neurologic manifestations. Ocular lesions are common and may include the following: 1. enlarged lacrimal glands (rare), 2. fragmentation or segmentation of the blood column in the conjunctival vessels, 3. chronic iridocyclitis and secondary glaucoma, and 4. hemorrhages in the retina with or without a central exudative detachment of the retina. The details of 20 cases of the syndrome are tabulated. Two-thirds of these cases showed the

hemorrhages and central detachment noted above. The author therefore feels that this combination is pathognomonic for the disease. These findings are of serious prognostic import. All patients studied by the author died in two to three years. There is no known treatment. (10 figures, 1 table, 15 references)

David Shoch.

Falls, H. H., Schlaegel, T. F., Jr., Harley, R. D. and Snydacker, D. **Symposium: Psychosomatic ophthalmology.** Tr. Fifth Pan-Am. Cong. Ophth. 1:427-457, 1956.

Falls, Harold H. **The role of inheritance in psychosomatic ophthalmology.** pp. 427-432.

Constitution in biology is defined as the sum total of the individual's hereditary make-up and of his past and present experiences. When the eye patients are seen under this light, one is practicing psychosomatic ophthalmology—a most successful approach. Genetic factors determine in certain siblings the same type of reaction under similar stress. Adult and juvenile glaucoma follows an irregular dominant pattern of inheritance. Hydrophthalmos shows considerable evidence for autosomal recessive inheritance. Axenfeld's syndrome and aniridia are dominantly transmitted. In refractive errors genetic prognosis should be determined by individual analysis of each family since hereditary changes are sometimes dominant, sometimes recessive. In hysterical amblyopia, which is diagnosed by exclusion of organic disease, heredity has a major role but the specific type of inheritance pattern is unknown. Finally in strabismus, the role of emotions is well known and a dominant mode of inheritance is common to convergent and divergent strabismus.

Schlaegel, T. F., Jr. **Vision.** pp. 433-440.

Refraction is a field rich in psychosomatic problems. Asthenopia can be due not only to refractive errors but to other factors such as hypochondriasis, frustration at the near task, fear of blindness, extraorbital muscle tension, and decreased level of emotional wellbeing. Almost all the above symptoms can be relieved by a change of glasses, by a change of environment or by simple and well-aimed reassurance. True hysterical amaurosis is rare. In a serial study of 800 patients of the Eye Clinic of the Indiana University Medical Center, 5 percent had tubular hysterical fields, but only two of the 40 patients had demonstrable transient amaurosis. (9 references)

Harley, R. D. **Some psychosomatic aspects of primary glaucoma.** pp. 441-448.

The close relationship between emotional disturbances and glaucoma is well known. The hypothalamic regulation of these mechanisms is beyond voluntary control and thus far removed from preventive therapeutic measures. Further evidence has been added for the autonomic nervous system control of the intraocular vascular circulation, secretion of intraocular fluid and level of ocular tension. Five case histories illustrate these facts. There is a question whether in chronic simple glaucoma there is a similar mechanism. In certain cases tranquilizers, as adjuvant therapy, have proved highly effective. (15 references)

Snydacker, D. **Psychosomatic aspects of concomittant heterotropia.** pp. 449-452.

The author believes that an emotional imbalance may actually result in a heterotropia, in contrast to the traditional sequence of ocular imbalance leading to emotional disturbances. The diagnosis is difficult and requires individual analysis, no general rules can be laid out. Obviously, glasses are of no avail since refractive error has been previously ruled out. The majority of these cases are nonaccommodative esotropias. Surgical treatment is doomed to failure in those cases that appear to be surgical but are merely

functional. If no expert psychotherapist is available one should abstain from offering therapy. It is the author's experience that attempted therapy may be detrimental. In general the prognosis is poor unless there is a radical change in the factors which have produced the disturbance.

Snydacker, D. **Neurocirculatory disturbances.** pp. 453-457.

Central angiospastic retinopathy is considered a functional neurovascular response to stress. Different individuals react in a dissimilar manner to the same stressing agent. Heredity has not been shown to play a specific role in this condition. Therapy consists in adequately informing the patient about the psychosomatic ethiology of his condition, restriction of caffeine and tobacco, the prescription of thiamine hydrochloride, nicotinic acid and mamitol hexanitrate three times a day in addition to intravenous thyroid H antigen three times a week. Papaverine and Diamox have proved useful. Steroids are not used by the author since they themselves are stressful agents. In constitutional epilepsy there is a very close link, as demonstrated by E.E.G. studies, with paroxismal cerebral dysrhythmia. This dysrhythmia is apparently gene-controlled and is most likely inherited as a dominant trait. The prescription of dilantin and phenobarbital, and reassurance to the patient are accepted methods of therapy.

Migraine very often attacks the driving, tense, anxious, energy-consuming and hyperactive patient. In the female the close relation to menstrual periods suggest an abnormality of the electrolite metabolism. Heredity plays a definite role and the disorder is believed to be transmitted by a dominant gene.

Gustavo Scioville-Samper.

Fedatove, T. A. **Visible vessels of the eye and of the retina in coarctation of the aorta.** Vestnik oftal. **2**:3-12. March-April, 1958.

Between 1955 and 1957 the author studied six patients with coarctation of the aorta of whom two were operated upon for relief of the coarctation. The retinal arteries showed notable narrowing, tortuosity and pulsation. Retinal findings ordinarily noted in other hypertensive conditions were completely absent. Diastolic and systolic pressures in the retinal artery as measured with the aid of the dynamometer were found to reach a level of almost twice normal. Ocular tension was always within normal limits. Fluctuations in the size of the blind spot were observed and apparently depended on the state of the circulatory supply. Blood vessels of the conjunctiva were markedly tortuous. After successful elimination of the coarctation in one patient the systemic blood pressure dropped sharply, while systolic and diastolic pressures in the central retinal artery dropped moderately. (2 figures, 1 graph, 1 table, 8 references) Victor Goodside.

Healy, James J. **The intra-ocular circulation in arteriosclerosis and high blood-pressure.** Tr. Ophth. Soc. U. Kingdom **77**:3-30, 1957.

The author emphasizes the fact that the ophthalmologic aspects of a general disease cannot be accurately interpreted without some knowledge of the changes proceeding elsewhere in the body. Senile arteriosclerosis, manifest by hyperplasia of the medial coat of the arteries in the aging, is common, but rarely commences before the age of 50 years. The relative height of the systolic to the diastolic pressure is important. The fundus may show the arteries reduced in caliber irregularly and their light reflex broadened and dull; one branch of an artery may become pale and associated with hemorrhages. The veins may be veiled and at any arteriovenous crossing there may be

partial constriction with banking. Exudates may be: 1. white, sharp-edged, 2. fluffy white fibrinous (cotton wool), and 3. grayish white and peripapillary. Hemorrhages may be 1. aneurysmal or petechial, 2. superficial and flame shaped, and 3. they may be sub-hyaloid.

Treatment must be applied to the hypertensive patient only after careful study and prolonged assessment of the individual case and subject to flexible readjustment throughout life. (7 tables, 45 references) Beulah Cushman.

Lijo Pavia, J. **Retinal arterial pressure in pregnant patients.** Rev. oto-neuro-oftal. **33**:36-38, April-Aug., 1958.

The author emphasizes the importance of ophthalmo-dynamometry in pregnant patients and its significance when it is important to know the value of the pressure in the cranial portion of the carotid artery. After studying a large number of patients, he reached the conclusion that the diastolic pressure in the central retinal artery does not change appreciably during the last days of pregnancy and the first few days after delivery. He feels that this study should be carried out in all patients in whom preeclampsia is suspected. (8 figures) Walter Mayer.

Lopez Garcia, Eloy. **Collagen diseases in ophthalmology.** Arch. Soc. oftal. hispano.-am. **18**:375-384, May, 1958.

The etiology, pathogenesis, gross and microscopic pathology, blood changes and response to therapy of collagen diseases is reviewed. The ocular complications of collagen diseases are discussed, and it is concluded that they have no definite etiologic, physiopathologic, anatomic or therapeutic characteristics. The author believes that the concept of collagen disease is transitory, and that with further scientific advances the clinical picture which it comprises will be more precisely classified. Ray K. Daily.

Mavioglu, Hilmi. **Behçet's recurrent disease: analytical review of the literature.** Missouri Med. **55**:1209-1222, Nov., 1958.

In this extensive review the author points out that the lesions of the eye are the gravest manifestations of Behçet's syndrome. Its cause is a virus. Complete blindness almost always occurs and there is no known therapy. (1 figure, 185 references) F. H. Haessler.

Scott, G. I., Ashton, N., Nabarro, J. D. N. and Lister, J. **Discussion: Ocular aspects of diabetes.** Tr. Ophth. Soc. U. Kingdom **77**:115-158, 1957.

Scott, G. I. **Ocular aspects of diabetes.** pp. 115-126.

In his introductory contribution to a discussion of the ocular aspects of diabetes Scott limits his remarks to retinopathy. He points out that it can occur in the absence of arteriolar hypertension or arteriolar sclerosis. The incidence increases with the duration of the disease. It has been suggested that the retinal proliferation which occurs as a complication of diabetes is essentially proliferation of vascularized tissue secondary to vitreous hemorrhage. There is a question, however, whether the hemorrhage causes the vascularization or is itself the result of rupture of new-formed vessels. The author thinks there are probably two types of diabetic retinopathy. He doubts that the retinopathy is the result of an inherited vascular defect capable of autonomous expression not dependent on a disturbance of carbohydrate metabolism. He thinks that there is ample evidence that diabetic retinopathy is the result of a defect of metabolism but that it concerns more than just carbohydrate metabolism. (6 figures, 13 references)

Ashton, Norman. **Experimental aspects of diabetic micro-angiopathy.** pp. 127-140.

Characteristic vascular complications of diabetes are called diabetic microangi-

opathy. Symptoms of renal disease occurring in diabetes can be hypothetically explained as the result of a seepage of lipoproteins and mucopolysaccharides from the circulation through foci of capillary degeneration. Retinal micro-aneurysms affect the venous side of the capillary network, in contrast to the renal nodules which are arterial lesions and occur only in diabetes. Fat emboli are common in the diabetic group and may be responsible for capillary lesions. Retinal and renal lesions are closely related to an elevation in the blood of lipoproteins, and there is a close relation between these lesions and elevation of the mucopolysaccharides in the blood.

If a true experimental analogue of diabetic glomerulosclerosis can be produced without retinopathy, then we can suppose that diabetic retinopathy, at least in the early stages, may have a different histogenesis and should be treated as a separate experimental problem. (4 figures, 2 tables, 60 references)

Nabarro, J. D. N. **Diabetic retinitis.** pp. 141-149.

Diabetic retinitis varies with the duration of the diabetes and the age of onset. It has been reported in 77 percent of 159 patients who had diabetes more than 15 years and it appeared five to 22 years after the diabetes was diagnosed. Joslin has reported the retinopathy in 49 percent of 451 patients who had developed diabetes before they were 30 years old. In diabetes there is a complex biochemical disturbance of protein and fat metabolism as well as carbohydrate utilization. Adrenal cortical activity probably plays a part in patients with retinopathy but adrenalectomy has proved disappointing in attempts to stop retinitis.

There are insulin-deficient diabetics with low plasma insulin who are usually thin and under 30 years of age, sensitive to exogenous insulin, and liable to diabetic ketosis. There are also insulin-resistant diabetics who are usually obese and over 50 years of age. The two types require different treatment which the author outlines in this essay. (2 figures, 17 references)

Lister, J. **Diabetic retinitis.** pp. 151-158.

The author reports a study of 100 patients with diabetes and eye complications. He discusses cataract and retinopathy as manifestations of diabetes and refers briefly to iritis, blurring of vision, glaucoma, ocular nerve palsies, homonymous hemianopsia, blepharitis, optic atrophy and retrobulbar neuritis. He emphasizes the fact that retinopathy and cataract are the most common ophthalmic manifestations of diabetes and points out that every patient with cataract should have an examination of the urine.

Beulah Cushman.

Sheehan, B., Harriman, D. G. F. and Bradshaw, J. P. P. **Polyarteritis nodosa with ophthalmic and neurological complications.** A.M.A. Arch. Ophth. **60**:537-547, Oct., 1958.

In this very extensive case report the general and neurologic findings, and the severe ocular complications in one eye in a 35-year-old woman with polyarteritis nodosa are described. (6 figures, 25 references) G. S. Tyner.

Siegert, P. **Giant cell arteritis as a cause of acute blindness.** Klin. Monatsbl. f. Augenh. **133**:417-419, 1958.

The author comments on the paper by Seitz, Klin. Mbl. f. Augenh. 132:383. He prefers the term "temporal arteritis" and believes that the excision of the temporal artery is of diagnostic, therapeutic and prophylactic value in these patients. (1 figure) Frederick C. Blodi.

19

CONGENITAL DEFORMITIES, HEREDITY

Amalrik, Jammes and Bessou. **The syndrome of Porak-Durante and the disease**

of Lobstein. Bull. Soc. belge d'opht. **118**: 336-350, 1958.

The syndromes of Porak-Durante and of Lobstein have been considered to be different manifestations of the same disease. Genetic studies and histopathologic analysis clarified common elements and distinguished individual clinical manifestations dependent on the date of onset and the course. Lobstein's disease is transmitted in dominant autosomal form. Pronounced fragility of bones, blue scleras, deformities of the skull and more or less complete deafness are the most significant signs and symptoms. The circumstances of survival are favorable as the general resistance increases, and the tendency towards fractures even decreases with age. Porak-Durante's disease seems clinically to belong to the osseous dystrophies. Severe bone deformations and multiple incompletely healed fractures are already visible at birth. Other distinctive features are dwarfism and failing cranial ossification. There are no characteristic ocular abnormalities. The prognosis of this type of osteogenesis imperfecta congenita is poor; death mostly occurs during infancy or childhood. The authors report two cases and review the literature. (11 figures, 24 references)

Alice R. Deutsch.

Appelmans, M., Michiels, J. and Doyen, N. **Congenital malformations and endocrinopathies of pregnancy.** Bull. Soc. belge d'opht. **118**:391-404, 1958.

In this retrospective study on mothers of 30 children with ocular malformations the findings were summarized in table form and included comments on accompanying somatic anomalies, on anomalies of brothers and sisters, also on ancestors and finally the results of the physical examinations of the mothers. In 20 out of the 30 mothers, hormonal disturbances were evident, a very significant finding for the ophthalmologist who realizes that adequate therapeutic preventive measures might be feasible in selected cases. Teratogenic data of diverse etiology are explained so that they can be understood and interpreted. Only a few cases could be ascribed to hereditary traits. The Rh factor was not responsible for a single ocular anomaly. There was also no case of hereditary syphilis. Maternal infectious diseases (one case of toxoplasmosis, two of German measles, one of influenza) were the cause in only four affected children. Diabetes and hypothyroidism of the mother were the main offenders with the most deleterious effects during the early period of pregnancy. The complexity of the endogenous and exogenous factors and their effect on organogenesis is extreme and in spite of the notable work already done, much more study will be necessary to clarify the role of the various pathogenic forces. (3 tables, 22 references)

Alice R. Deutsch.

Delmarcelle, Y. and Pivont, A. **Familial heterochromia associated with ocular and somatic malformations: syndrome of Waardenburg-Klein.** Bull. Soc. belge d'opht. **118**:380-391, 1958.

The characteristic signs and symptoms of Waardenburg's syndrome include: lateral displacement of the internal lid angle with elongation of the canaliculi and blepharophimosis, broadening and hyperplasia of the root of the nose, hyperplasia of the inner half of the eyebrows, partial or complete heterochromia of the iris, complete or incomplete deafness, and partial albinism, mostly in the form of isolated strands of white hair. The affliction is hereditary and the transmission is dominant with single or combined penetration of the diverse symptoms. This fact was very well documented by the first personal observation of the authors. They presented a pedigree of seven generations afflicted with this syndrome. In this particular family the abnormalities of the

eyebrows and the deformation of the root of the nose were the most persistent manifestations. Deafness and heterochromia were only seen occasionally. The second patient was a 22-year-old man who displayed the complete syndrome but had a negative family history. Therefore this could be interpreted as a sporadic case and as such demonstrates the possibility of mutation.

A recognition of the Waardenburg-Klein syndrome is of practical, clinical interest because it includes about 1.4 percent of the existing deaf-mutes and because carriers of very inconspicuous signs and symptoms could eventually have deaf-mutes among their descendants. (5 figures, 23 references)

Alice R. Deutsch.

Dollfuss, M.-A. **Another case of scleroderma "à coup de sabre" with many ocular lesions which developed during an observation period of 12 years.** Bull. Soc. belge d'opht. **118**:377-379, 1958.

The author describes the case of a 50-year-old woman who had been under observation for 12 years. She presented a band-shaped, partially hypertrophic type of scleroderma in the frontal region, initially without osseous changes but later with visible atrophic changes of the left frontal, parietal and sphenoidal bones. Vascular abnormalities were visible first in the right retina only, later also in the left. There were many recurrences of the neuroretinal disease, followed later by spells of iridocyclitis which were complicated by clouding of the lenses. Two years preceding her death a hemiatrophy of the left half of the face developed rapidly with pronounced enophthalmos and wasting of the left nostril. The patient died suddenly of a brain hemorrhage at the age of 50 years. The parents of the patient were first cousins. There was also a history of congenital syphilis for which she was extensively treated in younger years, but no relationship to the oculocutaneous disease could be established.

Alice R. Deutsch.

François, J. **A new syndrome.** Boll. d'ocul. **37**:162-194, March, 1958.

The author describes a syndrome which, he believes, represents a clearly defined clinical entity. It is characterized by birdhead dyscephalia with dyscrania, mandibular aplasia, dental anomalies, dwarfism, hypotrichosis, cutaneous atrophy, microphthalmia, and congenital cataract. (25 figures, 3 tables, 34 references)

Joseph E. Alfano.

François, J., Verriest, G., Rabaey, M. and De Rouck, A. **A new pedigree of mice with absent sensory retinal epithelium.** Ann. d'ocul. **191**:592-597, Aug., 1958.

The authors found several mice in a mixed litter of white mice who had no sensory retinal epithelium. They state that such animals can be recognized by their smaller than normal eyes. In three such animals the ERG was found to be absent and histologic study showed an absence of rods, external granular layer and external plexiform layer. The pigment epithelium was well preserved. The internal granular layer and the internal plexiform layer appear normal; however the number of ganglion cells is reduced. (4 figures, 8 references)

David Shoch.

Freeman, J. D. J. **Reading difficulties in childhood.** Tr. Ophth. Soc. U. Kingdom **77**:611-613, 1957.

In 1908 congenital word blindness was found to occur in one of every 2,000 London school children. In 1942 McMeekan found it in 10 percent of Edinburgh school children, with a definite familial tendency and more common in boys than in girls. The cause of the disability is unknown, but it may be a manifestation of developmental aphasia. (5 references)

Beulah Cushman.

Hermann, Pierre. **Syndrome of microphthalmos, retinitis pigmentosa, and glaucoma.** Arch. d'opht. **18**:17-24, 1958.

The author has studied a family in which in four generations, 13 of 17 subjects had this syndrome. He describes its essential features in detail and notes that in addition to the triad—microphthalmos, retinitis pigmentosa, and ocular hypertension—a number of other signs may be associated. Of these cataract is the most frequent. The syndrome can be transmitted through several generations but may appear also as a single example in a family. The author comments on the prognosis of the disease, noting its evolution to blindness. Surgical treatment of the glaucoma is usually unsuccessful. (1 figure, 6 references) P. Thygeson.

Lavergne, G. **Hypermetropia gravis and hereditary simple microphthalmia.** Bull. Soc. belge d'opht. **118**:350-356, 1958.

Microphthalmia is a rare hereditary anomaly. The mode of heredity varies, being either dominant with inconstant penetration or recessive. Two children, brother and sister among nine normal children, were presented. They showed the signs and symptoms of simple microphthalmos, namely reduced size of the cornea, increased corneal curvature, shallow anterior chamber, reduced length of the eyeball, mesodermal tissue in the chamber angle and aplasia of the macula. The presence of mesodermal tissue in the chamber angle and the absence of differentiation in the region of the macula are ascribed to an arrest of development at the end of the sixth fetal month. Therefore pattern of heredity is a more probable etiologic factor than phenocopy in spite of the fact that complicated cases of microphthalmos have been ascribed to hereditary syphilis or toxoplasmosis. Microphthalmia with coloboma of the iris, lens, and choroid, and microphthalmos with diverse intraocular malformations represent two other types of tetralogic microphthalmos. (3 figures, 3 tables, 12 references) Alice R. Deutsch.

Lundström, R. and Boström I.-L. **Rubella in pregnancy and the incidence of developmental anomalies of the eyes.** Acta ophth. **36**:782-788, 1958.

An epidemic of rubella was observed in Sweden in 1951-1952; 29 children whose mothers had the disease during the first trimester of pregnancy were examined. Two (7 percent) were found to have congenital cataract, and six (21 percent) chorioretinitis. No defects were found in 25 children whose mothers had rubella during the second trimester. One child whose mother had the disease in the third trimester had chorioretinitis but not of typical appearance. No defects were found in the eyes of 48 children whose mothers had been in contact with rubella without being infected, and in 30 controls. (3 tables, 2 references)

John J. Stern.

Meunier, A. and Toussaint, D. **Scleroderma "à coup de sabre" with fundus lesions.** Bull. Soc. belge d'opht. **118**:369-377, 1958.

Ophthalmologic findings in the circumscribed type of scleroderma are rare while many ophthalmologic abnormalities have been reported with the diffuse type. Fundus lesions have never been observed together with either the plaque, the band, or the dotted variety of the localized form. Hemiatrophy of the face, however, has been attributed to isolated facial lesions of scleroderma.

The case of a young girl is described. She suddenly developed a bandshaped paramedian sclerodermic lesion after a severe emotional upset. Her general physical examination was negative. A mild hypesthesia of the ipsilateral cornea, a mild inequality of the pupils, and a localized perivascular sheathing of the su-

perior temporal retinal vessels without any interference of vision in the eye corresponding with the skin-lesion, were found during a routine ophthalmologic examination. The pathogenesis of the disease is briefly reviewed, especially in view of a possible relationship with the collagen diseases. There is also a short summary of previously published cases. (6 figures, 8 references) Alice R. Deutsch.

Schmelzer, Hans. **A microform of Marfan's syndrome.** Klin. Monatsbl. f. Augenh. **133**:471-477, 1958.

A mother and daughter are described who had ectopic lenses in each eye. The fingers of both patients had elongated middle and end phalanges, but normal first phalanges. (3 figures, 13 references)

Frederick C. Blodi.

Zanen, J. and Meunier, A. **Disparity in color perception in identical twins.** Bull. Soc. belge d'opht. **118**:356-368, 1958.

Disparity in color perception in identical twins is very difficult to explain. Only two similar cases have been observed and published. (Nettleship, 1912, and Walls and Mathew, 1952). The twins under discussion were females. Physically they were identical, except that the protanopic twin had a supplementary toe, a fact which might raise doubt of their univitelline origin. Therefore detailed clinical and laboratory investigations were made to establish the possible identity of the twins. Their history also revealed that only one placenta was found at their birth. Many tests were made to verify the type of dyschromatopsia which finally was diagnosed as protanopy. The father of the girls was also protanopic. The mother had normal color perception. There are two possible explanations for this case; both of them, however, are not quite satisfactory. The twins could be fraternal after all, in spite of the clinical tests in favor of their identity, or the protanopic sister may have presented the phenotypic manifestation of a heterozygotic stage. (6 figures, 9 references)

Alice R. Deutsch.

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HYGIENE, SOCIOLOGY, EDUCATION, AND HISTORY

Hunter, R. A. and Rose, F. C. **Robert Boyle's "uncommon observations about vitiated sight," (London, 1688).** Brit. J. Ophth. **42**:726-731, Dec., 1958.

"Boyle's law" is known to all students, but few know that he was created Doctor of Medicine of Oxford University in 1665 and that among his many writings are discussions of medical subjects some of which are related to visual disturbances. These writings are in reality detailed reports of his findings in patients with visual complaints. It is interesting to note that Boyle contributed to ophthalmology two classic descriptions dealing with exophthalmic ophthalmoplegia and dark adaptation approximately 200 years earlier than the writings of those to whom credit is given for the first description. (1 figure, 21 references)

Lawrence L. Garner.

Rodger, F. C. **The pathogenesis of ocular onchocerciasis.** Tr. Ophth. Soc. U. Kingdom **77**:267-289, 1957.

The author presents the problem of blindness of a half a million people in Africa between latitude 20°N and 10° south of the equator. The individual density figures (I.D.F.) for onchocerciasis are an index of the degree of infestation of the individual. The cornea and iris were involved only if the I.D.F. was high, usually over 25. (12 figures, 2 tables, 19 references) Beulah Cushman.

NEWS ITEMS

EDITED BY DONALD J. LYLE, M.D.

411 Oak Street, Cincinnati 19, Ohio

News items should reach the editor by the 10th of the month. For adequate publicity, notices of postgraduate courses and meetings should be received three months in advance.

ANNOUNCEMENTS

SOUTH-EAST METROPOLITAN REGIONAL HOSPITAL BOARD CORNEO-PLASTIC CONFERENCE: A PRELIMINARY NOTICE

A conference will be held on June 26 and June 27, 1959, at the Queen Victoria Hospital, East Grinstead, Sussex. There will be a symposium on "Problems of tissue transplantation" and lectures on surgery of the lids, cornea, lacrimal apparatus, and socket. Illustrative cases, operations on closed-circuit television, and scientific exhibits will be shown.

The conference will be strictly limited to 60 members, including places reserved for visitors from abroad and for nonconsultant surgeons.

An inaugural dinner will be given to members and their wives on June 26, 1959. Excursions have been arranged to Canterbury and Petworth for the ladies.

Registration fees. Consultants, £5 5s. Od. Nonconsultants, £3 3s. Od. For further information please apply to the Secretary, Corneo-Plastic Unit, Queen Victoria Hospital, East Grinstead, Sussex.

STANFORD POSTGRADUATE CONFERENCE

Stanford University School of Medicine will present the annual postgraduate conference in ophthalmology from Monday, April 6, through Friday, April 10, 1959. Registration will be open to physicians who limit their practice to the treatment of diseases of the eye, or the eye, ear, nose, and throat. In order to allow free discussion by members of the conference, registration will be limited to 30 physicians.

Instructors will be Dr. Dohrmann K. Pischel, Dr. Jerome Bettman, Dr. Max Fine, Dr. Earle H. McBain, and Dr. Arthur Jampolsky.

Programs and further information may be obtained from the Office of the Dean, Stanford University School of Medicine, 2398 Sacramento Street, San Francisco 15, California.

SEMINAR ON GLAUCOMA

A seminar on glaucoma with particular emphasis on gonioscopy and the study of the anterior angle will be given at the Brooklyn Eye and Ear Hospital on May 18, 19, and 20, 1959. Ample opportunity for practical instruction in the use of the gonioprism will be given and material from the glaucoma clinic will be utilized.

The course will be given by Dr. Daniel Kravitz, assisted by Drs. Mortimer A. Lasky, Nicholas P. Tantillo, and Samuel Zane. Registration is limited to six ophthalmologists only.

Application and the fee of $50.00 may be addressed to: Dr. Daniel Kravitz, Brooklyn Eye and Ear Hospital, 29 Greene Avenue, Brooklyn 38, New York.

POSTGRADUATE COURSES

The Institute of Ophthalmology of the Americas, New York Eye and Ear Infirmary, will offer the following postgraduate courses:

1. "Ocular surgery," May 18 to 29, 1959. It will include cataracts, keratectomies and keratoplasties, pterygium, lacrimal sac, retinal detachment, muscles including ptosis, glaucoma, enucleation and evisceration, and orbitotomy. Registration is limited. Preference will be given ophthalmologists taking the entire course. If places are available, doctors may register for individual parts.

2. "Histopathology of the eye," May 18 to 23, 1959. Limited registration.

For further information, please write: Mrs. Tamar Weber, Registrar, Institute of Ophthalmology of the Americas, New York Eye and Ear Infirmary, 218 Second Avenue, New York 3, New York.

PAN-PACIFIC SURGICAL ASSOCIATION

The eighth congress of the Pan-Pacific Surgical Association will be held in Honolulu, Hawaii, September 28 through October 5, 1960.

All members of the profession are cordially invited to attend and are urged to make arrangements as soon as possible if they wish to be assured of adequate facilities.

An outstanding scientific program by leading surgeons promises to be of interest to all doctors. Nine surgical specialty sections are held simultaneously.

Further information and bronchures may be obtained by writing to Dr. F. J. Pinkerton, Director General of the Pan-Pacific Surgical Association, Suite 230, Alexander Young Building, Honolulu 13, Hawaii.

COURSE ON LIGHT COAGULATION

Prof. Dr. Meyer-Schwickerath will give an introductory course on light coagulation from May 4th to 8th. The course will be limited to those individuals who have the opportunity to work with light coagulation, or who expect to have such opportunity in the near future. For further information write

to Prof. Dr. Meyer-Schwickerath, Bonn-Venusberg, Universitäts Augenklinik.

Brooklyn Eye and Ear Hospital

The eighth annual scientific session sponsored by the Brooklyn Eye and Ear Hospital Alumni Association will be held on April 4th at the Brooklyn Eye and Ear Hospital, 29 Greene Avenue, Brooklyn. Taking part in the ophthalmology program will be Dr. Walter V. Moore, Dr. Edwin N. Beery, Dr. Dan M. Gordon, Dr. Robert Trotter, Dr. Carteret Lawrence, Dr. Richard C. Troutman, Dr. Daniel Kravitz, and Dr. Mortimer A. Lasky. There will be a panel discussion of glaucoma and Dr. Troutman will present a preliminary report on the "Use of alpha-chymotrypsin in ophthalmology." It is requested that all problems or questions for panel discussion be sent to Dr. Abraham M. Sands, 874 Carroll Street, Brooklyn 15, New York.

Czechoslovakia meeting

The Ophthalmologic Section of the Czechoslovakia Scientific Medical Society Jan Evangelista Purkyně will meet on September 25th and 26th at the Spa Luhačovice. A symposium on "Electroretinography" is being planned for this meeting and it is requested that anyone wishing to participate in this symposium communicate with Prof. Dr. Jan Vanýsek, Pekařska 53, Brno, Cazechoslovakia, not later than May 1st. The symposium will be preceded by a conference on "The pathology and therapy of diseases of the lens." Those who wish to present papers at the meeting should submit a brief abstract to Dr. Vanýsek before April 1st.

Oregon meeting

Dr. Walter S. Atkinson, Watertown, New York, and Dr. Robert N. Shaffer, San Francisco, will be guest speakers at the 18th annual ophthalmology and otolaryngology postgraduate convention sponsored jointly by the Oregon Academy of Ophthalmology and Otolaryngology and the University of Oregon Medical School. The meeting will be held June 26th to 30th during the Oregon Centennial Exposition and International Trade Fair. For further information write to Dr. Robert E. Fischer, 1216 S.W. Yamhill Street, Portland 5, Oregon.

Films for Pan-American meeting

Any person in North America having films on ophthalmologic subjects who would desire to show them at the Pan-American Congress of Ophthalmology in Venezuela, January 31 to February 7, 1960, should contact either Dr. Wendell L. Hughes, 131 Fulton Avenue, Hempstead, New York, or Dr. Louis J. Girard, Hermann Professional Building, Houston 25, Texas, as soon as possible.

Persons living in South America who wish to present films should get in touch with Dr. Alejandro Salleras of Buenos Aires, or Dr. O. Velasquez of Panama, or Dr. R. Valenzuela of Santiago, Chile.

The films should be new films which have not been shown elsewhere. A summary of the film with the title should be sent, along with data as to whether it is silent or sound (optical or magnetic), and the length of showing time, and a summary of the subject matter.

Miscellaneous

Symposium on retinal diseases and surgery

At the ninth annual session of the New Orleans Academy of Ophthalmology, held in New Orleans February 23rd to 27th, a symposium on "Diseases and surgery of the retina" was presented;. Guest speakers were Dr. Arthur J. Bedell, Albany, New York; Dr. Harold F. Falls, Ann Arbor, Michigan; Dr. Bertha A. Klien, Chicago; Dr. Dohrmann K. Pischel, San Francisco; Dr. C. Wilbur Rucker, Rochester, Minnesota; Dr. Charles L. Schepens, Boston; Dr. Donald M. Shafer, New York; and Dr. Lorenz E. Zimmerman, Washington, D.C.

Virginia research laboratory

The Elbyrne G. Gill Eye and Ear Foundation announces the establishment of a research laboratory in conjunction with the Eye-Bank and Sight Conservation Society of Virginia under the direction of Miss Jean Swartz, M.S. in biochemistry. The laboratory is prepared to make the dye test for toxoplasmosis.

Societies

New Mexico meeting

Dr. Peter C. Kronfeld, Chicago, will be the guest speaker at the annual meeting of the New Mexico Ophthalmological Society to be held in Santa Fe on April 30th and May 1st and 2nd. Dr. Kronfeld will speak on "Complications of cataract surgery," "Current techniques of retinal detachment surgery," and "Diagnostic problems." Ophthalmologists from seven Rocky Mountain states are being invited to the meeting, and the invitation is extended to all ophthalmologists who wish to attend.

Brooklyn meeting

At the 151st regular meeting of the Brooklyn Ophthalmological Society held at the Brooklyn Eye and Ear Hospital, Dr. Daniel Kravitz acted as moderator of the symposium on "Glaucoma." Panelists were Dr. Alphonse A. Cinotti, Dr. Bernard Kronenberg, Dr. Bernard Schwartz, and Dr. Richard C. Troutman.

American College of Surgeons

Presiding over the ophthalmic surgery program of the four-day sectional meeting of the American College of Surgeons held at Saint Louis, Missouri, March 9th to 12th, was Dr. Robert D. Mattis, Saint Louis. A symposium on "Ophthalmic surgery of trauma," was presented at the first session. Taking part in this were Dr. Erwin E. Grossman, Milwaukee; Dr. Merrill J. Reeh, Portland, Oregon; Dr. Phillip Ellis, Little Rock; and Dr. Ichiro D. Okamura, Boston. Taking part in the second symposium on "Ophthalmic surgical infections," were Dr. Henry F. Allen, Boston; Dr. William B. Clark, New Orleans; Dr. W. Howard Morrison, Omaha;

and Dr. Albert N. Lemoine, Jr., Kansas City, Missouri.

American Goiter Association

The 1959 meeting of the American Goiter Association will be held April 30th and May 1st and 2nd at the Drake Hotel, Chicago. The program for the three-day meeting will consist of papers and discussions dealing with the thyroid gland, its physiology, pharmacology, pathology, and therapy.

Puget Sound Academy

The Puget Sound Academy of Ophthalmology and Otolaryngology held its annual president's program Friday and Saturday, January 9th and 10th, at the Rainier Club in Seattle. Guest speakers were:

Dr. Robert S. Pollack, San Francisco, who spoke on "Neck tumor diagnosis," "Tumor surgery of the orbit," and "Head and neck tumor surgery."

Dr. Aram Glorig, Jr., Los Angeles, who presented the method for computation of hearing disability newly approved by the Committee on Conservation of Hearing of the American Academy of Ophthalmology and Otolaryngology, and played a tape recording of how the English language probably sounds to persons with certain types of hearing impairment.

At the Saturday evening meeting Dr. Edison G. Dorland, Seattle, outgoing president of the academy, presented a gavel to Dr. Willard F. Goff, Seattle, the new president. Dr. George H. Drumheller, Everett, is president-elect. Dr. James L. Hargiss, Seattle, was re-elected secretary-treasurer. Dr. H. Fred Thorlakson and Dr. Robert A. Campbell, both of Seattle, were elected new trustees.

Certificates of Fellowship were presented to the following doctors voted fellows during 1958: Robert E. L. Shumate, Seattle; Lester T. Jones (honorary) Portland, Oregon; Phillip A. Peter, Seattle; Ferris F. Ketcham, Seattle; William P. Berard, Seattle; and Cornell E. Blackham, Seattle.

Southern officers

At the recent meeting of the Southern Medical Association the following officers were elected for the Section on Ophthalmology and Otolaryngology:

Dr. G. S. Fitz-Hugh, chairman, 104 East Market Street, Charlottesville, Virginia; Dr. George M. Haik, chairman-elect, 812 Maison Blanche Building, New Orleans 16, Louisiana; Dr. B. Russell Burke, vice-chairman, Suite 147, 490 Peachtree Street, N.E., Atlanta 8, Georgia; Dr. Mercer G. Lynch, M.D., Secretary, 3503 Prytania Street, New Orleans 15, Louisiana.

The next meeting of the section will be held in Atlanta, Georgia, from November 16 to 19, 1959. Those interested in participating should write to the secretary of the section, Dr. Lynch.

International Society for Clinical Electroretinography

During the International Congress of Ophthalmology held in Brussels in September, 1958, an International Society for Clinical Electroretinography (ISCERG) was organized. The objective of the society is to promote the work in clinical electroretinography by preparing and conducting meetings and symposia, by establishing a central bibliography distributed regularly to the members, and by suggesting a standardization of the instrumentation and procedure in clinical electroretinography and of the recording and measurement of clinical electroretinograms.

The work of the society will be carried out by a Central Board of Officers, a General Committee, and an Advisory Board.

The officers of the society are as follows: President, G. Karpe (Stockholm); vice-presidents: A. Franceschetti (Geneva) and J. François (Gent); secretary general: H. E. Henkes (180 Schiedamsevest, Rotterdam, Netherlands); secretary for the Western Hemisphere: H. M. Burian (Department of Ophthalmology, University Hospitals, Iowa City, Iowa); treasurer: W. Straub (Universitäts-Augenklinik, Hamburg 20, Germany).

The membership fee has been set at 10.—DM (approximately U.S. $2.50) a year, payable in DM or local currency.

Anyone residing in the Western Hemisphere and interested in this new society is requested to contact Dr. Burian. All others should apply to Dr. Henkes.

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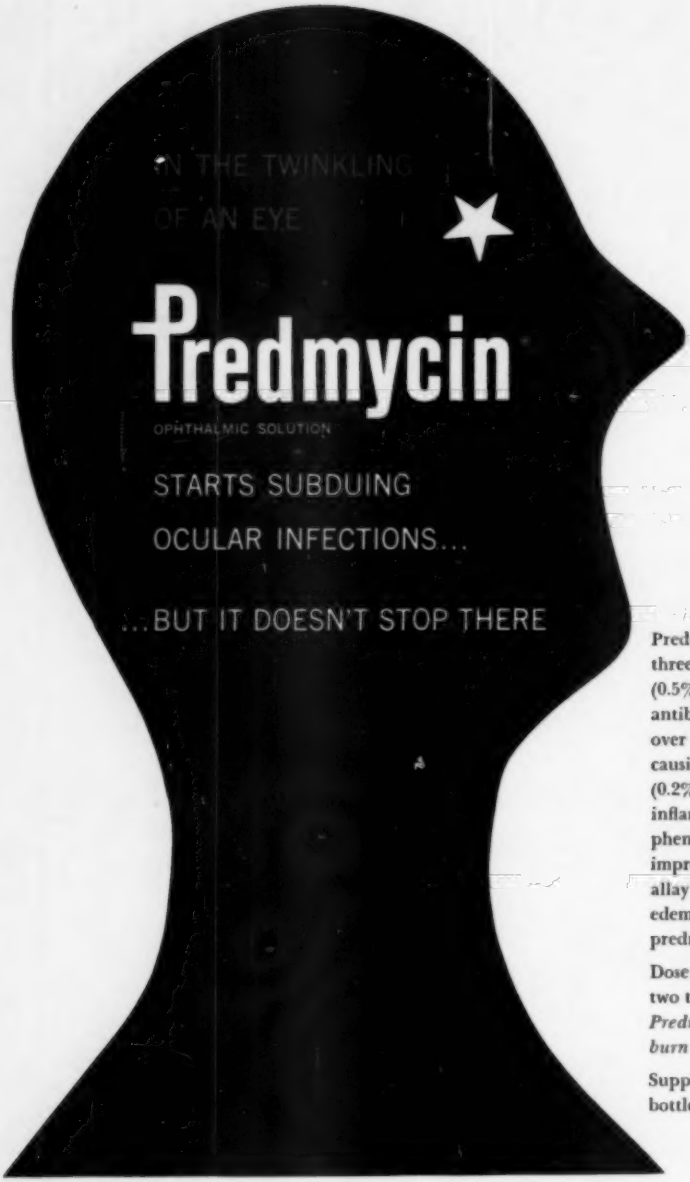

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